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THE SO-CALLED UNIT CHARACTERS IN RELATION TO HEREDITARY DISTURBANCES OF THE NERVOUS SYSTEM*

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NEW YORK

The primary purpose of this Association is research into the problems of neurology and psychiatry. Therefore no attempt is made at a meeting to cover a subject in its entirety; only new phases and new aspects of disease are presented, with so much of what is already known as will serve for the necessary background. The remainder of the subject—that which is generally accepted—forms no part of our discussions and deliberations, but may appear in concise form in our annual volume.

Our present venture is into the realm of heredity. The "origin of species" has given us an understandable hypothesis of the gradual mutations, the reasons for survival and for extinction, the necessity for over or under accentuation of parts, color, form, habits, which make up the characteristics of species and their differentiation from one another—so far as our finite mind can encompass the intricacies of a law of our infinite universe, whose beginning and ending are unknown.

The origin of the individual is the problem of our century. Why are not all members of a species alike? What underlies their differences of appearance, of reactions, of ability; of their differences in the sphere of biochemistry, of their differences in their powers of immunity to disease? How much is the individual the mere integration of a highly complex mathematical equation whose terms and factors and coefficients were introduced by all his ancestors—and how will he himself change the terms of this equation—if at all—the integration of which will be again another individual? What does he obtain from the past, what does he give to the future? He, as a complexity of units, may be studied as a link between the origin of life and its destination. If he is studied as a link, then we must know him in his relation to the adjoining links—that is to say, his relation to his ascendants as well as to his descendants—a consideration often missed by investigators who will look on him as an end-product.

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* Presidential address read at the Association for Research in Nervous and Mental Diseases.

So our facts bearing on the individual's life history in all its phases must be gleaned from his children as well as from his parents. And these facts, the basic ones, those which technically in the study of inheritance are known as "unit factors," from the specific material with which our laws of inheritance have been built.

The difficulty which immediately confronts us is the determination as to which characters are "unit." Let us take several characteristics which have frequently been made the subject of investigation as to their transmissibility by inheritance on a mendelian basis—abnormal growth in height, migraine, epilepsy and diabetes. Are these really unit factors, or are they but evolved from unit factors through environmental or other influences?

First: Is growth in height a unit character? Much work has been done to determine the law of the transmission of this so-called "unit character." The conclusions derived from many exhaustive investigations are not definite as to the existence of such a law on a mendelian basis in the human species.

Second: It has been a favorite study of those interested in heredity and eugenics to determine the transmissibility of epilepsy, as though this were akin to a "unit character." There are as many types of epilepsy as there are of cough, and their bases of origin are just as widely different in the former as in the latter. It would be just as proper to discuss a table of heredity of "cough" as one of epilepsy.

Third: Migraine is another so-called "unit character" in disease, and no textbook on nervous and mental diseases is complete without a table on inheritance of migraine. And yet types of migraine are seemingly well defined and recognized as differing among themselves, just as do the types of epilepsy. But, as with epilepsy, these types are all grouped under the generic term "migraine."

A fourth extremely interesting characteristic which seems to occur in family groups is disturbance of the blood sugar. Many of the persons with this disturbance, because of the spilling over into the urine of the sugar, have been called diabetic.

Just as interesting, although not as yet sufficiently observed, is the group with low blood-sugar content. This, so far as I know, has not been grouped as an inheritable characteristic, although from observations made on a few hundred persons, it seems to bear the same relationship to some underlying transmissible factor as does high blood-sugar content. The individual manifestations of low blood-sugar are of extreme importance, and it is these manifestations that have served to connect in my mind heretofore seemingly isolated disease processes described as inheritable, all of which are probably but end results of a real basic unit factor and not the unit character in themselves.

Let us scan the four manifestations just presented: abnormal growth, epilepsy, migraine and disturbed blood-sugar balance. Epilepsy and migraine have for some decades been recognized as having some relationship to each other, but just what that connection is has not been understood. It was seen that both occurred in the same families, were inheritable and occasionally occurred in the same person as mutually substitutive. In my service, for some years we have been studying migraine, and one interesting observation made is that preceding an attack of certain types of migraine, the blood-sugar is below normal. Preceding many of the so-called idiopathic epileptic attacks also the blood-sugar is low, and all recent observers of the effect of insulin in diabetes report the appearance of convulsive seizures when the blood-sugar drops to a critically low level. Again, in our statistics covering rapid and great growth in adolescents—including many hundred cases—we so frequently obtain a history of migraine not only in the family and antecedents of the patient, but even in himself during the period of growth, that the position of the simultaneous occurrence of migraine and the rapid growth being coincidental cannot be maintained. Hence, these four conditions—epilepsy, excessive growth, migraine and blood-sugar disturbance—seemingly as far apart as the poles, have possibly some interrelated factors controlling all and undergoing, in the various members of a family or in successive families, metamorphosis in its manner of appearance. If this be so, then the metamorphosed end product is not that with which we should be working in the study of the hereditary transmission, but this ought to be the underlying factor itself. So the history of our patient with migraine should not only contain the antecedent sufferers from this complaint alone, but should include also those that had epilepsy, extreme growth, or blood-sugar disturbances as well. Of course the blood-sugar disturbances are difficult to get at, especially as the low-content group are not as yet in a definite symptomatic class and hence are not recognized as such. And so, for some years in our clinic we have insisted that our histories of migraine should contain direct references to antecedent epilepsy, excessive growth, and blood-sugar disturbances when these can be obtained, either in ascendants or descendants, and a surprising mass of material is gradually being accumulated pointing to an hereditary underlying factor associated with or even controlling these end products. This underlying factor is primarily an anatomic one, we believe, or at least the indications point that way, and possibly consists in a conformation of the base of the skull by which the sella turcica is crowded and overhung by its protective bony capsule, making the pituitary body incapable of fully performing its metabolic work at first, while later, owing to the inherent tendency of living tissue toward physiologic

activity, it makes way for itself through erosion by pressure within this capsule. Then as a result of such pressure, migrainous attacks supervene, blood-sugar disturbances are produced, and with them occasional convulsive seizures occur, while the increase in pituitary activity synchronously determines increased body growth. In all probability, many other end-products of this original unit factor will come to be likewise associated with these, such as a disturbed fat metabolism, gonadal abnormalities and behavioristic and other mental manifestations. Such an anatomic characteristic can much better be understood as being a unit factor in inheritance than can the complicated biochemical processes leading to the superficial pictures which we see. Indeed, if the latter could be considered as unit characters, then a human zygote would have to possess many more than forty-seven or forty-eight chromosomes, which at present are allowed him, or it. One can readily see from the processes above mentioned—even though he may not bring himself to accept the specific examples cited—how a simple anatomic trait or property, depending on its mere physical situation, can be fraught with the most diverse clinical dynamic results. If the anatomic characteristic were such an obvious one as that of polydactylism or syndactylism, or of single muscle absence, it would be comparatively simple to construct tables showing the laws of its transmission—as is done; but when an entire disease process is under discussion, then the feat of inheritance table construction and even more so, its interpretation, is extremely difficult, for we never can be certain that the disease process is not merely one end-product of many arising from the same transmissible simple factor as yet undiscovered. Such a theory would practically make any hereditary transmissible diseases dependent on constitutional hereditary factors; the individual's environment and training and the demands made on his resources determining the particular channel in which such an hereditary factor would develop its manifestations. The difference in individuals from a common stock would then depend not only on the difference in the unit characters of constitution transmitted from the parents, but also on the development from such units via different pathways of end-products of appearance, behavior, or so-called disease.

One can readily see that those individuals in whom transmitted unit characters develop in lives at variance with their environmental necessities, must have great difficulty in survival, while those in whom these developments are in harmony with environmental demands remain to procreate and continue the stock. Sir Frederick Mott, in his Maudsley Lecture of 1921, states the matter as far as mental diseases are concerned, as follows: "Mental diseases tend to terminate in three generations; the stock dies out or changes to more normal types." In the

former alternative he says that in each succeeding generation, the mental disease appears earlier in life and hence procreation is gradually eliminated, thus stamping out the disease; while in the latter, he simply begs the whole question of inheritance of unit characters. Moreover, his theory is not in accord with our studies in this country—witness the Kalikuk and Juke families! That is to say, the stock is not improved, probably, by a change in transmissible unit characters—which change is said to be impossible—but by a change in the development of these unit characters through education, training and biochemical means to environmentally harmonious end-products. So I think it most important that we recognize first what are unit characters, and that we do not confuse them with their offspring, appearance, behavior and disease.

When it comes to the inheritability of a highly complex disease, such as for example progressive muscular dystrophy, another famous subject of inheritance tables, we are in all probability again dealing with a combination of several unit characters all of which must be present in order that an end-product of such complexity may arise. For, be it observed, this disease is not one of the muscular system alone, but includes in the dystrophic process, the bony skeletal framework of the body, the heart and cardiovascular system, the genital system, some of the cell groups of the central nervous system and the biochemical and endocrine systems. It is practically universal in its attack. Now we find individuals—and often in dystrophic families—who have many of the dystrophic signs elsewhere than in the muscle system, and hence without the characteristic outstanding disability. These individuals are not classified in the disease tabulation—but they do have at least some of the characteristics of the disease—and hence probably one or more, but not sufficient, unit factors are operative in them. An understanding of these unit factors would give us real information about the disease and its inheritance and its variability in the individual it attacks—all of which is missed when the disease process per se is regarded as a unit disturbance.

And so I desire to point out our great fallibility in considering disease processes—especially constitutional ones as distinguished from extraneous and accidental ones—as unit processes with well defined boundaries. They are certainly not that. And our study of the heredity and transmissibility of these diseases must be made on a foundation of their basic unit factors and not on the finished disease process as we see it. These basic units will probably be found in simple anatomic, physiologic and biochemical variations, which when expanded by the proper combinations and permutations will produce all of our transmissible hereditary diseases in their full panoply of symptoms.

I trust, in conclusion, that I may have made clear the necessity, in studying heredity in disease, to determine not merely the existence in ascendant or descendant of the similar set of symptoms connoted by this disease in the patient, but further, to consider the possibility of frequently recurring constitutional anomalies in the family studied as having some bearing on the pathogenesis of the disease in question and perhaps even of such an anomaly concealing one of the true unit factors of the disease.

HEREDITARY FACTORS RESPONSIBLE FOR DEVELOPMENT OF OPTIC ATROPHY AND RETINITIS PIGMENTOSA*

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The hereditary conditions underlying the two degenerative processes —optic atrophy and retinitis pigmentosa—are similar to a certain extent. Leber's disease is not common; many ophthalmologists of wide experience have never with certainty seen a case. No microscopic examination has been made in this condition so far as I know. The primary pathologic manifestation of Leber's disease is in the form of an axial rather than a peripheral degeneration. The papillomacular bundle is vulnerable; and it is very sensitive to various environmental poisons, bacteriologic and chemical—quite apart from the effect of an hereditary taint. In the reports of institutions, it is not always possible to separate this type of optic nerve involvement so as to yield data of scientific value; and unless the family history is accurate, the factor of heredity is uncertain. Its time of onset is usually about the twentieth year, although authentic cases beginning at 5 and 6 years of age have been reported. The diagnoses of cases occurring after the age of 60 are open to suspicion on account of the many sources of retrobulbar neuritis and atrophy furnished by the toxemias, focal infections and disturbances of the pituitary body. Most of the cases occur in males, the taint being handed down through the mother. Leber gives the proportion of females affected as 10 per cent.

In this affection, as in pigmentary degeneration of the retina, the interesting feature is the inheritance of tissue vulnerability, making the optic nerve fibers in this disease especially susceptible to extraneous conditions and poisons. Consanguinity does not seem to be a factor.

RETINITIS PIGMENTOSA

In pigmentary degeneration of the retina, there is a general agreement about the ultimate pathologic condition present, but there is a difference of opinion in regard to the sequence of events. All experienced observers agree that this disease shows no evidence of inflammatory phenomena. Therefore, the term "retinitis pigmentosa" is a misnomer and should be abandoned. One might use the designation, "pigmentary degeneration of the retina;" or Collins' suggestion of

* Read at the Fourth Annual Meeting of the Association for Research in Nervous and Mental Diseases, New York, December, 1923.

"abiotrophy of the retinal neuro-epithelium," or the term "retinal abiotrophy," which combines brevity with a sufficiently accurate description of the condition.

Pathology of the Disease.—For a number of years, two views have been held concerning the pathology of this disease. The first attributes the defect to some condition of the optic nerve, particularly that set of fibers which run to the equatorial zone of the retina (Graefe, Leber, Lister and others). The second ascribes the degeneration to the special arrangement of the blood vessels of the anterior layers of the choroid and the posterior layers of the retina in this intermediate zone, which makes them more liable to disease or partial occlusion (Gonin, Nettleship and others). However, it is agreed that later there is an atrophic condition of the nervous elements of the retina, hyperplasia of the connective tissue and an advancement of the pigment cells, or their granules, into the vacant retinal spaces. This may be due to the functional capacity of these cells to advance toward the light; or the accumulation of pigment cells may be due to their mechanical occupation of spaces left vacant by absorption of the nervous structures of the retina. The slit lamp reveals in other conditions the wandering tendency exhibited by the pigment cells of the retina and the iris. Later, there is atrophy of the optic nerve and a tendency to the formation of a posterior cortical cataract. Collins—following Gowers' explanation of degenerative changes in certain hereditary diseases of the nervous system—believes that the process is one of primary degeneration of the neuro-epithelium, later with atrophy of the nerve fibers and an increase in the fibrous tissue elements. He therefore feels that it should be classed as an abiotrophy. I am inclined to believe with Collins that the degeneration is a truly abiotic process. Certain it is that sclerosis of the choroidal vessels does not produce a picture identical with this disease, and in some microscopically examined cases the choroid has approximated the normal. There are reported six cases of pigmentary degeneration of the retina in which such an examination of the specimen has been made. Knape's case occurred in an elderly man (exact age not given). The patient was myopic. A detached retina, together with iridocyclitis, necessitated the enucleation of the eye. The ages given in four of the other five cases were 35, 60, 65 and 67. For a more satisfactory establishment of the exact pathologic changes, it is unfortunate that—so far as I am aware—no microscopic examination has been possible in the early stages of the disease in an uncomplicated case. It is felt that such specimens would more certainly exhibit the abiotic nature of this affliction.

Practically all writers state that this affection is a bilateral disease. However, as careful an investigator as Nettleship states: "In the cases that follow, there can be no doubt that the disease was anatomically the

same as common binocular retinitis pigmentosa." He then reports, including the one case of his own, nine definite cases of monocular pigmentary degeneration of the retina. He rejects two other previously recorded cases. He notes the fact that in not one of these monocular cases was there a family history of the disease. He accounts for the possibility of these cases "by supposing that the vital endowments of the choroid are sometimes different at birth in the two eyes." Lutz says that we have to eliminate the assertion that hereditary diseases of the eyes are always developed bilaterally. In addition to other cases of unilateral inheritance, he quotes six cases of monocular color blindness, one of them occurring in a woman. Cases in which only one eye is affected must indeed be rare when observers of the greatest experience state that the condition is always bilateral. But we must accept Nettleship's statement that in rare instances it is entirely monocular. With his vast experience and his deep special interest in the subject, it is not likely that his case, or the ones he accepts, were instances of mistaken diagnosis. I have never seen a case in which only one eye was affected. But I have, on several occasions, seen cases in their incipiency which had been wrongly diagnosed as syphilitic choroiditis, and monocular chorioretinitis diagnosed as pigmentary degeneration of the retina. The occasional asymmetrical inheritance in these cases is somewhat emphasized, because the departure from the usual in transmitted conditions may possibly throw some light on these problems of heredity.

Incidence Among Males and Females.—Generally, experienced observers have found that more males than females suffer from this trouble. Nettleship gives the proportion as 60 to 40, which agrees with my experience. In Shoemaker's series of seventeen cases, only four were males—an example of these group coincidents that so often occur, although not in agreement with the whole mass of evidence. Nettleship finds in regard to hereditary sex transmission, that the proportion for the father is to the mother as 36 to 50.

Age Incidence.—In regard to the age at which this disease is recognizable: I have seen a well advanced case of pigmentary degeneration of the retina in a healthy girl of 8 months in a family in which several members were similarly affected. In a case recently observed in a healthy woman aged 45, with a good family history, good health and a negative Wassermann test, the trouble with the eyes was not noticed until after 25 years of age. The census report of 1920 stated that three persons had been blind from the disease at birth. In short, it may exist from the time of birth; it may come on shortly afterward; or, by reason of good general health, freedom from exciting causes, slight tissue liability, etc., it may not become evident until adult life.

Association With Other Conditions.—According to observers generally, this lesion is often associated with other physical or neurologic

stigmas. According to Nettleship, "It is estimated that at least 3.3 per cent. of persons with retinitis pigmentosa may be deaf, and that at least 4 per cent. of deaf mutes may have retinitis pigmentosa." Shoemaker says that "true retinitis pigmentosa is seldom if ever found in an otherwise perfectly normal and standard individual." Yet Nettleship (Case 1) reports that in a family history of seven generations, thirty-eight

TABLE 1.—*Institutions for Mental Defectives and Feeble-Minded*

Name of Institution	Number Inmates	Optic Atrophy				Examination	Remarks
		R. P.	H.	Not H.	Cong.		
Northern Wisconsin College and Training School	4,500*	0	2	0	0	Eye examination made	Present number 1,100
Michigan Home and Training School	2,330	0	0	0	0	Eye examination made	
Columbus (O.) Institution for Feeble-Minded	3,894	0	0	0	0	Eye examina-2 tion made	
Syracuse State Sch. for Mental Defectives	856	..	..	..	..	Physical exam-'nation made	Lack of staff
Rome State School, New York	2,776	..	..	..	..	Physical examination made	
Pennsylvania Training School	914	2	0	0	0	Eye examination made	
Exeter School, Rhode Island	.....	..	..	..	..	Not thorough physical examination	Lack of staff
New Jersey Institution for Feeble-Minded	.....	0	..	..	..	Eye examination made	
New Hampshire Sch. for Feeble-Minded	456	1?	0	0	0	Eye examination made	One case reported by oculist to be R. P.
Polk State School, Pennsylvania	2,100	1	0	0	0	Eye examination made	
North Dakota Institution for Feeble-Minded	386	..	..	..	..	No facilities for examination	Medically understaffed
Massachusetts Institution for Feeble-Minded	1,540	1	..	..	..	Eye examination made	Three optic atrophy, can not say heredity
Newark (N. Y.) State School	1,200	..	..	..	..	No eye examination made	
Indiana School for Feeble-Minded Youth	1,423	0	0	0	0	Eye examination made	None for several years
Lincoln State School and Colony	2,184	0	0	0	0	Physical examination made	Annual reports do not show any R. P. or O. A.
Letchworth Village, New York	1,729	..	..	..	..	No special eye examination made	
Totals.....	26,288	5 or 0.019%	2				

Total number in which eye examinations were made, 22,973 or 0.021%.

* While present number of inmates is 1,100, there have been 4,500 since the two cases of optic atrophy occurred.

persons were affected out of a total of 200, the males being slightly in excess. Then he adds: "Not a single case of deaf mutism, mental defect, or other degeneracy has occurred in this genealogy . . . . the ones seen were above the average of their class (artisans) in intelligence. They are decidedly long lived." In Snell's (Case 2) history of five generations, in which twenty-nine persons were affected out of a total of seventy-two, he says: "There appear to have been no cases of

deaf mutism, indeed, no degeneracy except that of the retina." While the percentages vary, most writers state that this disease is to be found in from 4 to 10 per cent. of deaf mutes, and not infrequently among idiots, epileptic persons, and others. Shoemaker reported seventeen cases of this affliction among pupils of a deaf and dumb institution. Usher found from thirteen to fifteen mentally affected out of sixty-nine patients. From the available data, it is certain that this affliction is often associated with other stigmas. But my personal experience leads me to believe that it may be found more often in otherwise normal persons than one would infer from the literature on the subject. In

TABLE 2.—*Institutions for Deaf and Blind, Deaf and Dumb and Deaf* *

Name of Institution	Number Inmates	Optic Atrophy				Examination	Remarks
		R. P.	H.	Not H.	Cong.		
South Carolina Sch. for Deaf and Blind	.....	..	..	..	..	.....	No information
New York Institute for Deaf and Dumb	450	1	0	0	0	Eye examination made	One case R. P. fifteen years ago
West Virginia Sch. for Deaf and Blind	268	..	..	..	..	No eye examination	Depend on history given by parents, etc.
Louisiana State Sch. for Deaf and Blind	.....	..	..	..	..	.....	No records
Mississippi School for Deaf	180	..	..	..	..	No eye examination	One optic atrophy, kind not stated
Pennsylvania Institution for Deaf and Blind	490	1?	0	0	0	No eye examination	One case several years ago
Indiana School for Deaf	349	0	0	0	0	.....	
Oregon State School for the Deaf	.....	0	0	0	0	.....	As far as known
Iowa State School for the Deaf	.....	0	..	..	..	.....	
Maryland State Sch. for the Deaf	152	0	0	0	0	Eye-examinations made periodically	
Nebraska State Sch. for the Deaf	.....	0	0	0	0	Eye examinations made when needed	
Totals.....	1,889	2 or 0.105%					

* All cases occurred several years ago; no case at present in a population of 782 where the eyes were examined. The total population since the occurrence of a case is greater.

other words, the degeneration approaches at times more nearly to the required mendelian "unit character." The very small percentage of this malady discovered in the institutions of this country is interesting; but one has to bear in mind the facts that conditions in private practice differ very much from those existing in public institutions; that the services of competent oculists are not often available; that in many cases a satisfactory ophthalmoscopic examination is impossible because of the fear manifested by the patient over such examinations.

There are certain points of interest brought out by Tables 1 and 2: (1) the great difficulty of getting not only good family histories, but accurate personal ones; (2) the great handicap that superintendents of institutions labor under from want of funds and lack of staff. This

scientific association can help to better these conditions. It is in these public institutions that we may hope to have the laboratory of the clinician for working out many practical biologic problems.

Frequency of Condition.—Dr. Loren Johnson writes that there have passed through his hands in the last four years about 400 cases of mental defectives, but that he does not recall having seen a case of retinitis pigmentosa among them. In my experience, the disease is often associated with high refractive errors, but during the last ten years, among

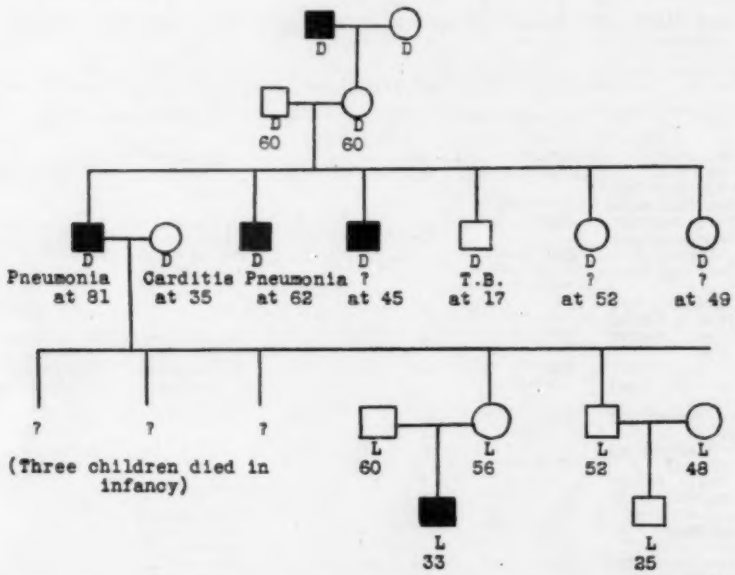

Fig. 1.—There is no family history of consanguinity, early deafness or other stigmas in this pedigree. There is a history of marked mental efficiency in the family. In the second generation, another unaffected daughter married the brother of her sister's husband, who was also unaffected. They had four children living to over 40 years of age. Two were boys and both were badly affected with pigmentary degeneration of the retina. The two daughters did not have the affliction, but passed it on to all of their male children. Squares indicate males and circles females. Solid black symbols indicate those affected; *D* indicates those dead; *L*, living.

a number of cases seen in private practice, only one patient exhibited any marked mental defect. This patient, a woman aged 37, was deaf from childhood, and her father reported that she was very nervous and resentful. Physical and other examinations were negative (including blood and spinal Wassermann tests). The patient was physically strong, and the disease was not recognized until the patient was 20 years of age. This case, too, is the only one I have seen in which there was practical blindness before 38 years of age. In contradistinction to this

case, a number of patients of the highest mental efficiency—writers, architects and others—have come under my care.

Typical Cases.—Within a week I have seen a case which is typical of a number that I have examined in private practice during the past thirty-four years. The man is 35 years old. His general health is excellent, with the exception of frequent colds and sinus involvement. To the latter infection may be attributed an ulcer of the left cornea in 1916. During childhood he experienced many diseases, cholera infantum, measles, typhoid fever, pneumonia and digestive disturbances. He made excellent records in his studies, and now occupies an important position in a large business which requires mental efficiency and considerable use of his eyes. He is fond of outdoor life, and during daylight drives his car himself. The sight of the right eye, with refraction error corrected, is 20/15-2. The sight of the left eye is reduced to 20/100, owing to the opacity of the cornea. Color sense is acute, light sense

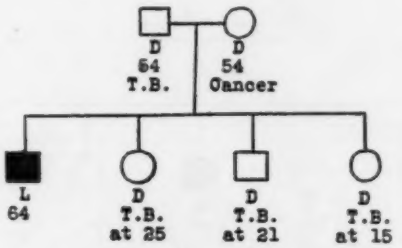

Fig. 2.—Pedigree of case recently seen by author.

much reduced, and the field of vision contracted to within 10 degrees of the point of fixation. His maternal grandfather, who had this disease, was a well-known country physician, who continued to practice and to drive himself everywhere until he was more than 50 years old. On the same side, an uncle, who is an unusually able physician, has sound eyes. He has been most helpful in his efforts to collect as many family facts as possible. The patient's pedigree—as far as it is obtainable—is given because it illustrates the fact that the unaffected daughter of an affected father transmitted the disease to her son. The meagerness of the pedigree shows how difficult it is to get a complete family history even in the case of the most intelligent people.

The pedigrees of two other recently seen cases are given.

In the first case (Fig. 2), the patient, aged 64, was the eldest of four children and the only living member of his family. In spite of cataract, he still has useful vision in a narrow field. The interesting feature of this history lies in the fact that of the five deaths in this family, one was from cancer (the mother), but the other four were from tuberculosis.

In the second case (Fig. 3) the patient was the youngest of five children. The family as a whole was long lived and healthy. One affected brother died of acute nephritis. The patient had cataract and no useful vision.

I have under observation at present three children with this defect of a family of four. The eldest child, a boy, showed this condition markedly at 14 months of age; the second child, a girl, was sound; the third, a boy, was affected at the age of 14 months, and the youngest child, a girl, at 8 months. As all of these cases were far advanced when the patients were first examined, they had evidently existed for some time—possibly from birth. At present, all of the children are rosy and well nourished, and mentally bright. The eldest boy is at the head of his classes. Psychologically, he would be classed with the group of "gifted children." All of the children have high refractive errors (far sight with astigmatism) and marked nystagmus. In addition, the oldest

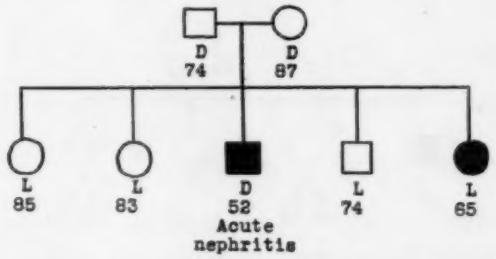

Fig. 3.—Pedigree of case recently seen by author.

child has an associated movement of the upper lid. The third child, a boy, was operated on at the age of 9 months for pyloric stenosis. His eyes grew much worse about the time of the operation. His case was the most advanced of the three. On neither side was there any family history of consanguinity or impaired vision. On the father's side, both the grandmother and her sister were deaf and afflicted with cancer, the sister in addition being mentally unstable. On this side, there had been a good deal of heavy drinking, on account of which the father himself was moderate in the use of alcohol. On the mother's side, there had been a good deal of deafness, but there was nothing else of importance in the family history. The cases of deafness were all noncongenital.

Racial Incidence.—This disease is stated to be most common in the Semitic race; but this does not coincide with my experience. While it does occur in the negro race, it is not common. Macnamara states that it is not infrequent among the Hindus, whose religion prohibits intermarriage, as many as twelve to fifteen cases a year having come under his observation. On the other hand, Kirkpatrick finds it common in

the South of India, where intermarriage takes place in 60 per cent. of the population—due partly to caste influence, and partly to the desire to keep family property intact. It is found among the Chinese and Japanese. I have never heard of a case in a North American Indian, although such cases might easily occur without coming under observation. Therefore, racial immunity to pigmentary degeneration of the retina does not seem to exist.

Prognosis.—The prognosis, as usually given, is unfavorable, the statement being made that blindness inevitably results. But in my experience, useful vision may continue for many years in spite of a gradually diminishing field of vision. I have seen several cases (includ-

TABLE 3.—*Census Report of 1920 on Blindness*

Total number reported		44
Male	31	
Female	13	
Native	42	
Foreign	2	
Negro	0	
Indian	0	
Age		
At birth	3	
Less than one year	1	
1 to 4 years	4	
5 to 9 years	2	
10 to 14 years	2	
15 to 19 years	2	
20 to 24 years	0	
25 to 34 years	4	
35 to 44 years	3	
45 to 54 years	12	
55 to 59 years	2	
60 to 64 years	2	
65 to 69 years	2	
70 to 74 years	1	
Total	40	
Age not definitely reported	4	
Parents first cousins	2	
Parents not first cousins	40	
(of these father blind 2, mother blind 2)		
Not reported	2	

ing the one reported by Harlan) in which in spite of old age, the patients were able—after the extraction of cataract—to do accurate close work. In this connection, it may be interesting to quote the following from the census report for 1920, which gives only forty-four cases of blindness from this malady:

The definition of "blindness" on which the report is based is found in the following instructions to be used in gathering the data:

> Include as *blind* any person who cannot see well enough to read, even with the aid of glasses. The test in case of infants must be whether they can apparently distinguish forms and objects; and in case of older persons who are illiterate whether they can presumably see well enough to read if they knew how to read. Do not include any person who is blind in one eye only.

While it is always possible that even with the greatest care some cases may not be reported, and that all cases may not be correctly diagnosed, the report is suggestive and interesting.

Consanguinity.—Consanguinity is given by the majority of writers the place next to heredity in the causation of the malady. Weeks states that "recent investigations prove that consanguinity may be regarded as the cause of about 25 per cent to 30 per cent of these cases." It happens that in the cases I have seen intermarriage has not been a potent factor. For instance, in the case of the three affected children, there is no history of consanguinity. On the other hand, I have had under observation for many years a woman in whose mother's family the history was negative. On her father's side, there was frequent intermarriage (three pairs of cousins); but at the same time, there was on this side a definite history of night blindness. The occurrence of this degeneration among the Hindus, who do not intermarry, has been mentioned. Nettleship says: "Though we are not yet able to disprove its specific influence, all the indirect evidence at our disposal points to marriage of blood relations being harmful only when both husband and wife are members of a tainted stock." According to Davenport: "It is not at all consanguinity that brings the trait out, but the increasing liability that consanguinity affords to the mating of two similarly defective germ cells." Herbert writes: "All that can be said on this question today is that where there is a taint in the family this is likely to be increased by the mating of two such affected individuals, while, on the other hand, with a healthy stock inbreeding seems to have no harmful effect whatever . . . . In short, consanguinity is harmful in tainted stocks, but harmless in healthy stocks."

COMMENT

A general survey of the subject allows one to sum up a few of the striking facts in connection with this malady. It is difficult to obtain histories of the afflicted members of the family. It is well nigh impossible to get the desired information concerning the unafflicted. The affection is a degenerative change, and there is strong evidence of its being a truly abiotic process. It is widely distributed, with no certain racial immunity. It appears at varying periods of life, in varying degrees of pathologic intensity and extent of tissue involvement. Syphilis does not seem to be a cause, although the two conditions may exist in the same person. The time of manifestation of this degeneration and its severity are influenced by the degree of the tissue liability, the general health of the subject and the occurrence of various superadded diseases like typhoid fever, pneumonia, syphilis, and exanthems, and other conditions.

In gathering data for consideration, one is impressed with the fact that in ophthalmic literature a clear and definite conception of what

heredity is frequently seems to be lacking. Davenport expresses what is clear from the eugenic standpoint when he states that it "is the phenomenon of the recurrence of traits in blood relations due to the persistence of their determiners in the germ plasm." And from a biologic standpoint, Menge's statement is explicit: "Whatever an offspring is to obtain from its parents must therefore be already present in the chromosomes of the germ cells of the parents or it cannot be inherited." The law of heredity, therefore, would rule out such statements as that syphilis is a hereditary disease due to the spirochetes of the mother infecting the fetus. The terms "congenital" and "hereditary" are too often, as pointed out by Howe, used interchangeably.

From a strictly biologic point of view, the Mendelian requirement of the involvement of a unit character is not as well satisfied by this disease as it is by some of the other hereditary eye conditions—notably congenital color blindness and lamellar cataract. Pigmentary degeneration of the retina is more complicated, and it is not always transmitted in the same way. In some instances, it is handed down by the father to both sons and daughters. In other cases it appears as a recessive and sex-linked degeneration. Occasionally, it seems to originate de novo, as if it were the result of some developmental abnormality. However, in this type, a complete family history might reveal the fact that it had merely been dormant for several generations, or that it was an equivalent inheritance of some other stigma. In my experience it has occurred at times in families in which many other members have suffered from tuberculosis (Fig. 2).

The statement made by Nettleship, in 1907, that it is the result of "tissue liability," still holds good. It would seem, therefore, to represent an ocular vulnerability rather than involvement of an inherited unit character. This vulnerability is much influenced by environmental conditions, both prenatal and postnatal. This tissue susceptibility has been noticed in other widely different somatic conditions. I have noted that in a family otherwise strong, healthy and long-lived, all the males for three generations were susceptible to poison ivy (*Rhus toxicodendron*), while all the females were immune. Only in the fourth generation did a female show any susceptibility to this toxic plant.

In spite of the epochal work of the biologist and the earnest help of the clinicians, our knowledge of heredity is still in its infancy. In regard to this retinal lesion, certain toxic substances, glandular, metabolic, chemical or bacterial, may play an important part in rendering these ocular tissues more susceptible to an untimely degeneration.

A meeting of such scientific and humane interest would not entirely fulfil its purpose unless a word were said concerning the cases that are brought to us for help. I am strongly opposed to the view—as sometimes given—that treatment is of no avail. It is well to remember, as

de Schweinitz so well states: "Good vision may last indefinitely in a narrow field." The general health should be built up. If syphilis coexists with this disease, it should have proper treatment. The same is true of anemia and faulty secretion of the endocrins. Focal infections should be removed. The refraction should be carefully made. Young children should have special instruction to overcome the ocular handicap. The therapeutic use of the roentgen ray has been advocated, and it is worth trying.

After all is said and done, the question arises, "How are these hereditary lesions to be prevented?" The most important step seems to be the realization of these conditions by the medical profession, and through the profession and students of genetics and eugenics, the education of the public in regard to marriage and the hereditary dangers.

In hereditary optic atrophy, a defective man should not marry, or at least have children. This is even more important in the case of an affected woman. But Davenport says: "that an unaffected female of an affected family may marry with impunity if all of her brothers are without defect and there are more than two of them." Of pigmentary degeneration of the retina, the same author states that "an affected man or woman should not marry even into stock without taint of retinitis." I feel strongly, when two normal persons produce an offspring with retinal degeneration, there should be no more children.

While pigmentary retinal degeneration is not responsible for a large percentage of loss of sight in the United States, the total amount of blindness from "hereditary ocular" lesions introduces a serious economic as well as humane factor for consideration.

Biologists tell us that "on the strictly scientific basis, everything that a man is, or can be, depends upon the factors of inheritance, environment, and training." The first factor is undoubtedly responsible for the tendency to an untimely death of the delicate cells, in the two ocular lesions we have considered. The eye is not surpassed by any other organ in affording an opportunity for the study of these dramatic incidents of local premature senility. It is an offshoot of the brain, and it is sensitive to any disturbances during its developmental period as well as to true hereditary taints. I dare to hope that ophthalmology may be able in the future to contribute its mite to the sum of knowledge of this potent force—heredity—which is as fascinating as life itself and almost as mysterious.

END RESULTS IN SIXTY-TWO CASES OF SPINA BIFIDA AND CEPHALOCELE*

GEORGE DAVID CUTLER, M.D.

BOSTON

The knowledge pertaining to spina bifida and allied conditions has been thoroughly presented by various authors of textbooks, notably Keen,[1] Pfaundler and Schlossmann,[2] Frazier,[3] Warbasse[4] and Abt.[5] In this community, valuable contributions to the subject have been made by Harmer,[6] and Woltman[7] has reviewed a series of 187 cases of spina bifida seen at the Mayo Clinic. Reference to the literature mentioned above will impress the reader with the seriousness of this malformation and the futility, in many instances, of attempting to effect a cure. The problem is a complicated one from many points of view. It is of interest to consider former methods of treatment. In this connection, it is worth while to read what Hilton[8] wrote, about the year 1860, concerning spina bifida and hydrocephalus. After a simple description of spina bifida, he pointed out the dangers associated with operations for this disease, especially those of injecting the sac with any irritating fluid. He also recognized the danger of meningitis following ligature. He advocated, as the safest and best method, aspiration to relieve tension and local support by application of a thick layer of collodion and by a bandage. Hilton noted that internal hydrocephalus was not an infrequent companion of spina bifida. In many cases of this disease, which he had examined after death, the cerebrospinal opening was closed, thus explaining how it happened that although the spina bifida had been tapped during life and no limit put

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* From the Surgical Service of the Children's Hospital.

* Read before the Boston Society of Psychiatry and Neurology, March 20, 1924.

1. Keen: Surgery, Philadelphia, W. B. Saunders Company **2**:820, 1912.

2. Pfaundler and Schlossmann (Trans. by Shaw and Fetra): The Diseases of Children, Philadelphia, J. B. Lippincott Company **5**:9, 1912.

3. Frazier: Surgery of the Spine and Spinal Cord, New York, D. Appleton & Company, 1918, p. 265.

4. Warbasse: Surgical Treatment, Philadelphia, W. B. Saunders Company **2**:349, 1918.

5. Abt: Pediatrics, Philadelphia, W. B. Saunders Company **2**:407, 1923.

6. Harmer, T. W.: Spina Bifida and Allied Malformations Based on an Operative Experience of Thirty-Four Cases, Boston M. & S. J. **177**:353, 1917; Spina Bifida—Operability and Time for Surgical Intervention, Ibid. **183**:775, 1920.

7. Woltman, M. W.: Minnesota Med. **4**:244, 1921.

8. Hilton, John: Rest and Pain, Ed. 5, New York, George Bell & Sons, 1892, pp. 33-40.

to the flow of fluid at operation; the patient did not suffer from cerebral congestion.

With the advent of aseptic surgery, the accepted method of treatment in suitable cases became excision of the sac with closure of the defect. Modern surgery thus made it possible to prevent deaths from rupture of the sac with leakage of cerebrospinal fluid and to decrease the deaths from meningitis but, on the other hand, may justly be accused of causing deaths by producing hydrocephalus. To operate or not to operate, and when, is the question, and the situation at times certainly presents a surgical dilemma.

It is the purpose in this paper to report the end results in fifty-seven patients with spina bifida and five patients with cephalocele, all but three of whom have been patients in the Surgical Service at the Children's Hospital, Boston.

TABLE 1.—*Results of Treatment of Spina Bifida in Children's Hospital, Boston, During Six Years*

Year	Admitted	Discharged Well or Relieved	Discharged Unrelieved	Discharged Untreated	Discharged to Other Departments or Hospitals	Dead	Remaining	Operations
1918	25	13	2	2	0	7	1	18
1919	21	8	4	2	0	5	2	13
1920	14	10	0	0	0	4	0	4
1921	13	9	0	0	0	4	0	9
1922	18	14	0	0	0	4	0	12
1923	13	4	0	0	1	7	1	9
Six years	104	58	6	4	1	31	4	65

The incidence of spina bifida is said to be one in every thousand births, but the Children's Hospital receives a good many of these patients for advice and treatment. Table 1 gives the number of patients brought to the hospital for treatment of this condition in the last six years.

Of 104 patients admitted, thirty-one died; a mortality of 29.8 per cent. Only sixty-five patients were treated by operations, with thirty-one deaths, a mortality not to exceed 47.69 per cent.

In addition to the patients admitted to the wards, the total number is raised by 122 [9] admitted only to the outpatient clinic, 16, 21, 18, 27, 18 and 22 in the six years 1918 to 1923, respectively.

These figures show the importance of spina bifida in this clinic. In addition to the patients from Greater Boston, many patients came from distant points.

The beginner is apt to feel that he can accomplish much by operation, but with the growth of personal experience and survey of results,

9. The majority of these patients are recommended to the hospital wards for treatment or are seen as postoperative, discharged patients from the wards, so that there is considerable duplication in the two sets of figures.

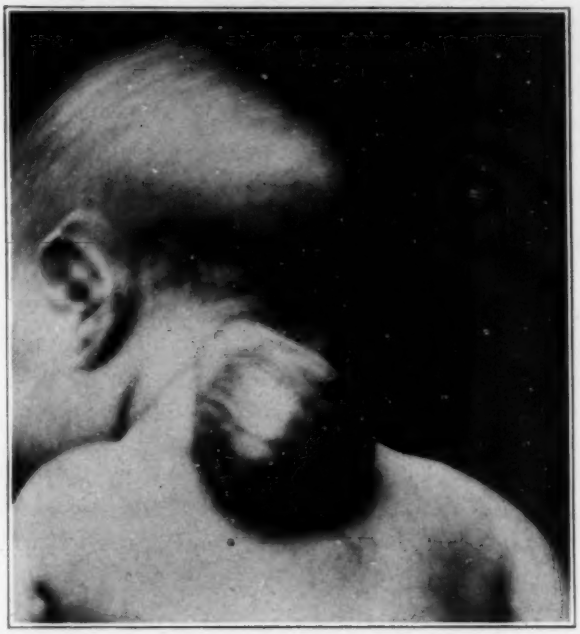

Fig. 1.—Operation series, Case 25, illustrating cervical pedunculated meningocele, favorable type for operation.

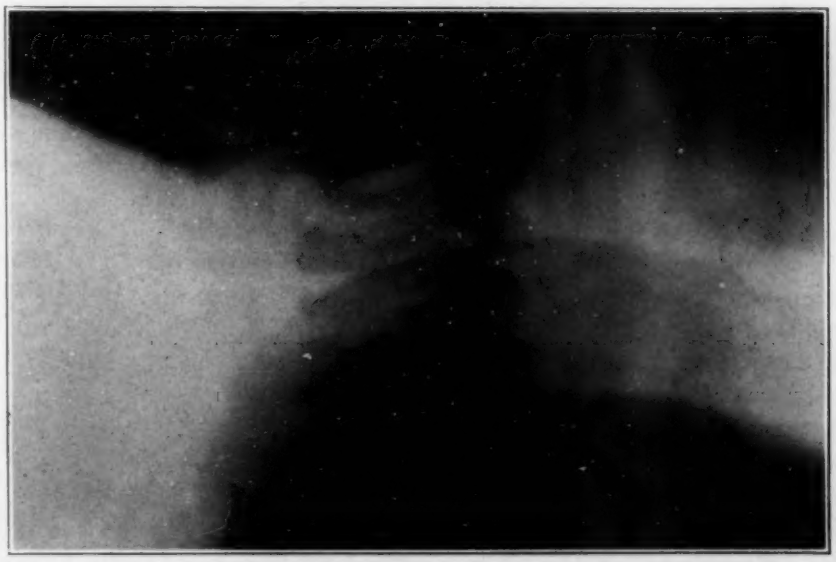

Fig. 2.—Roentgenogram of Case 25, cervical meningocele.

the ability to recognize the insuperable is attained. This has led me to recognize the dividing line, and I have, therefore, two groups of cases, those treated by operation and those regarded as inoperable. Unquestionably, the inoperable class should have included many which were subjected to operation earlier in the series.

It is of interest to note that Virchow ascribed the deformity to amniotic bands and local inflammatory processes. Von Recklinghausen thought disproportion between growth of the canal and the cord was responsible for it. Marchand and Ranke considered that imperfect separation of the skin and medulla caused the condition. The latter explanation is the most satisfactory. In addition to the faulty anatomic relation of tissues, we agree with Frazier and Harmer that the factor of increased cerebrospinal fluid pressure undoubtedly plays a part in the production and behavior of spina bifida. Clinical experience demonstrates the safety-valve action of the spina bifida sac.

It is easier to understand the distribution of this malformation if we remember that fusion of the spinal canal commences in the upper dorsal region and extends in both directions. Its failure causes a median posterior defect, which is most common at the lower part of the spine which closes last. In rare instances the protrusion takes place through a lateral or anterior defect. The defect may be confined to one arch, often it involves several, and rarely all. There are four general types of spina bifida: meningomyelocele, myelocystocele, meningocele and occulta.

CLINICAL MATERIAL

In my own series, there were thirty-nine patients with spina bifida treated by operation. The youngest was 18 hours old, and the oldest 3 years. Most of them were operated on before the age of 4 months; nineteen patients were less than 1 month old. The division according to sex was: twenty-six boys and twelve girls; in one case, the sex was not recorded.

Inquiry into the family history yielded positive information in only three instances. It was stated in one history that one other child had been born with spina bifida and in another that three previously born children had died—one of spina bifida and two of hydrocephalus—and a third parent wrote that the child born after the patient included in this series had spina bifida and died on the seventh day in convulsions. The important findings were: twenty-four meningomyeloceles and fifteen meningoceles; six cervical, four dorsal, nineteen lumbar, six lumbosacral and four sacral. Fifteen had coverings of good skin; twenty-four had thin membrane. Twenty-six of the sacs were unruptured; thirteen were ruptured or granulating. In twenty patients, the sac was of the raised type; ten were sessile and nine pedunculated. Four of the sacs presented a hemangiomatous appearance, and many

contained a considerable amount of adipose tissue. Twenty-three of the thirty-nine patients showed evidence of paralysis; in the remaining sixteen, it was not present or undetected. There was evidence of hydrocephalus in only nine patients.

OPERATION

The technic of operation followed was, to a certain extent, that used in the radical cure of hernia. Incisions were made through the skin and subcutaneous tissues, with isolation of the sac at its neck. A longitudinal incision was made through the side of the sac for exami-

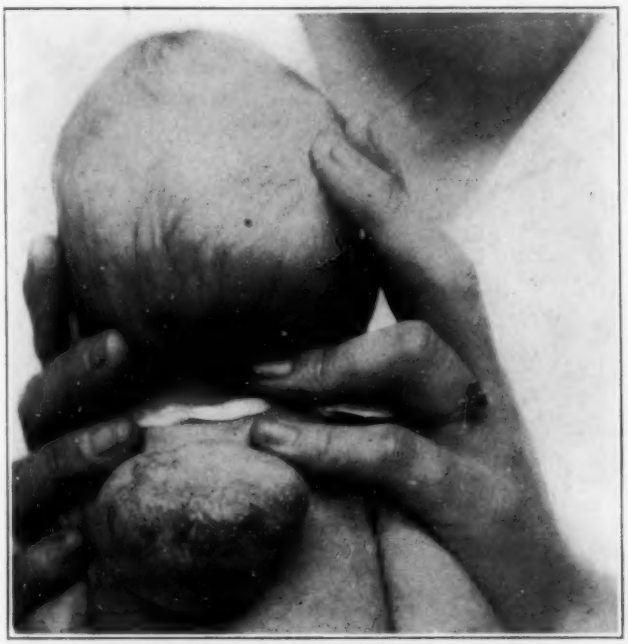

Fig. 3.—Operation series, Case 31; high dorsal meningocele, favorable type; excision—recovery—end result, well.

nation of its contents. Adherent nerve elements were freed and replaced in the spinal groove. The neck of the sac was sutured with continuous catgut, and the ends tied together if not too widely separated. The subcutaneous tissues were sutured together with interrupted stitches of fine plain catgut, and the skin edges approximated with interrupted stitches of fine silk. In twenty-five instances, no effort was made to reinforce this closure of the sac, because if tension is so great as to require extra support, hydrocephalus will probably develop and death ensue. In fourteen operations, lateral fascia flaps were used to support the sutured sac where the defect was large.

In the low spina bifidas, the long axis of the wound should be made in a transverse direction, as it keeps the wound higher and prevents soiling. I used it in twenty-four operations. Eleven of the incisions were vertical, one oblique and in three cases, in which a large circular excision of skin results, I used satisfactorily what I call the "Z" closure.

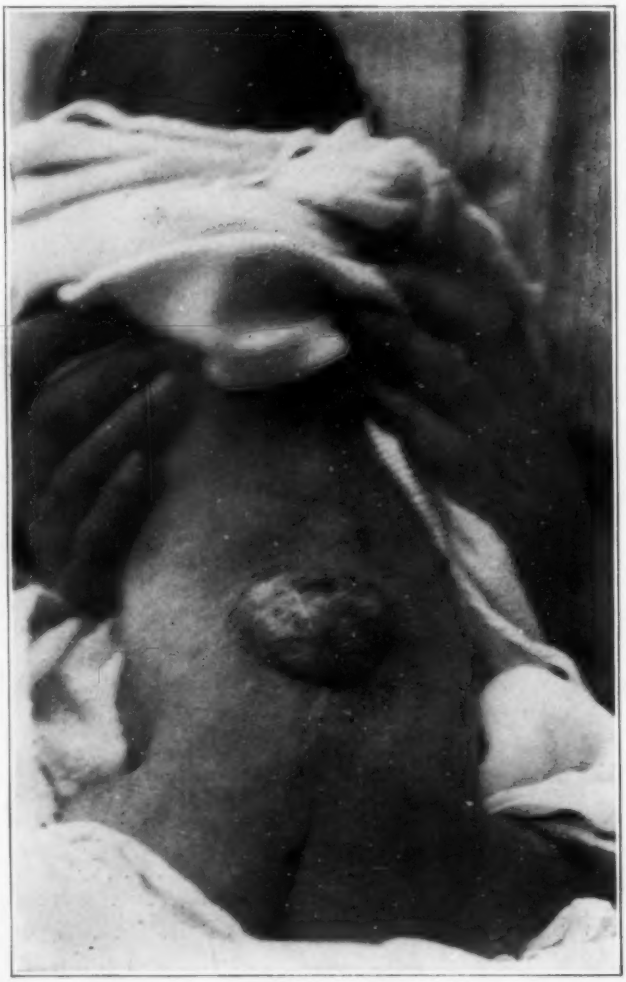

Fig. 4.—Operation series, Case 17, lumbar meningomyelocele of common type; hydrocephalus developed following operation.

In operations on these patients, the following procedures should be employed: (1) Bandage the extremities with flannel to retain heat. (2) Place the patient prone on one or two hot water bags under the thighs and lower abdomen to supply heat and elevate the buttocks.

(3) Ether is the anesthetic of choice which is started with the patient on the operating table and omitted at the earliest possible opportunity. (4) Careful hemostasis and speed in operation consistent with care in conserving essential structures; also gentleness in manipulation of the tissues to avoid shock should be carried out. (5) In tying off narrow

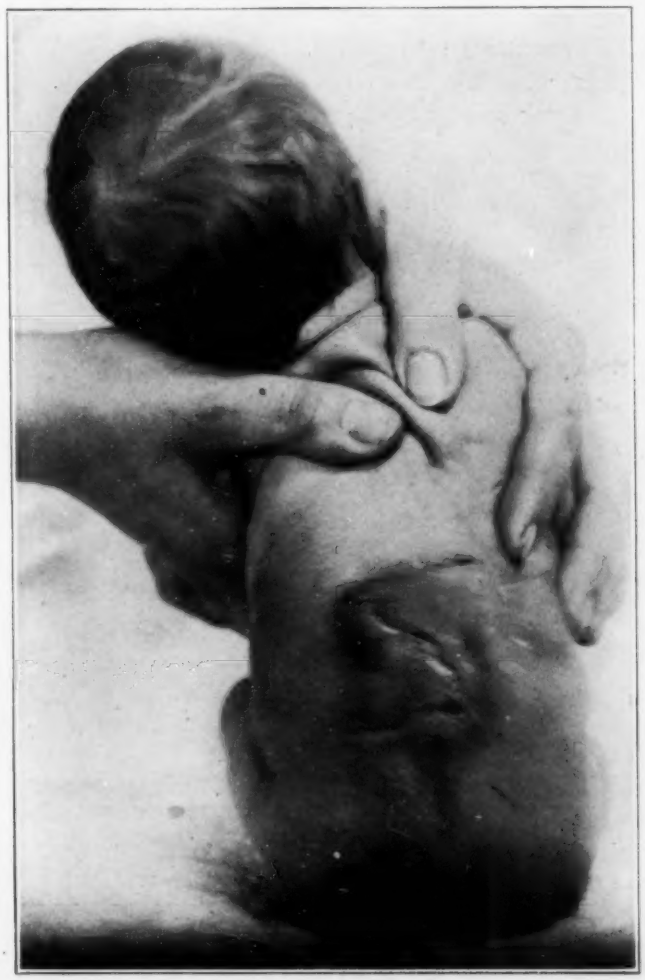

Fig. 5.—Operation series, Case 29, illustrating method of closing large, circular defect of skin. Patient had meningomyelocele. Operation was performed the day after birth. Hydrocephalus developed, and she died two weeks after leaving the hospital.

pedicles in order to avoid leakage from slipping ligature, transfix and tie, then place a tie just below the first one to prevent leakage through perforation. (6) Tension on the skin suture line should be avoided.

This may be relieved by lateral incision in the vertical closures and by incision above in the transverse closures. (7) A circular skin defect can be closed by lateral incisions above and below making two triangular flaps which slide by and when sutured make what I term the Z closure. (8) The gauze dressing is retained in position by imbricated strips of adhesive plaster.

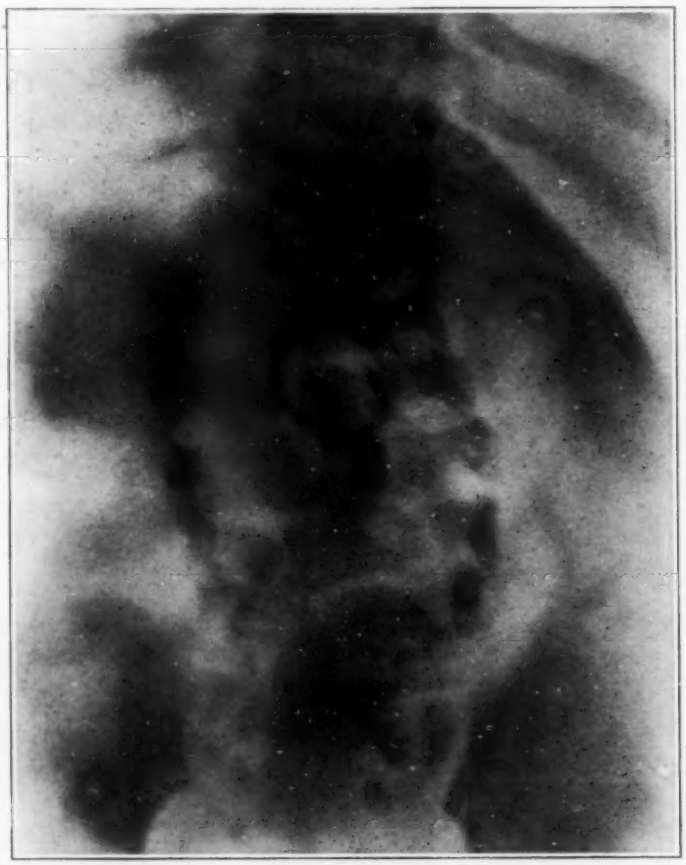

Fig. 6.—Roentgenogram of spine, no operation series, Case 10; taken when patient was 11 days old.

RESULTS

Twenty-two patients left the hospital alive (summary numbers 3, 5, 6, 9, 12, 16, 17, 18, 19, 20, 22, 23, 25, 26, 27, 28, 29, 30, 31, 32, 35, 37); sixteen died in the hospital.

CAUSE OF DEATH

Case 1.—Death occurred a few hours after the operation, the cause being doubtful.

Case 4.—Death occurred from acute hydrocephalus at the end of eleven days; the ventricle was tapped once.

CASE 7.—Death occurred from leakage of cerebrospinal fluid on the third day after operation.

CASE 8.—The temperature went to 107 F. on the day after operation, and the patient died of respiratory failure.

CASE 10.—Death occurred from hydrocephalus and meningitis two weeks after the operation (the sac was infected).

CASE 11.—Death occurred on the day after operation. Temperature, pulse and respiration were all elevated from unknown causes.

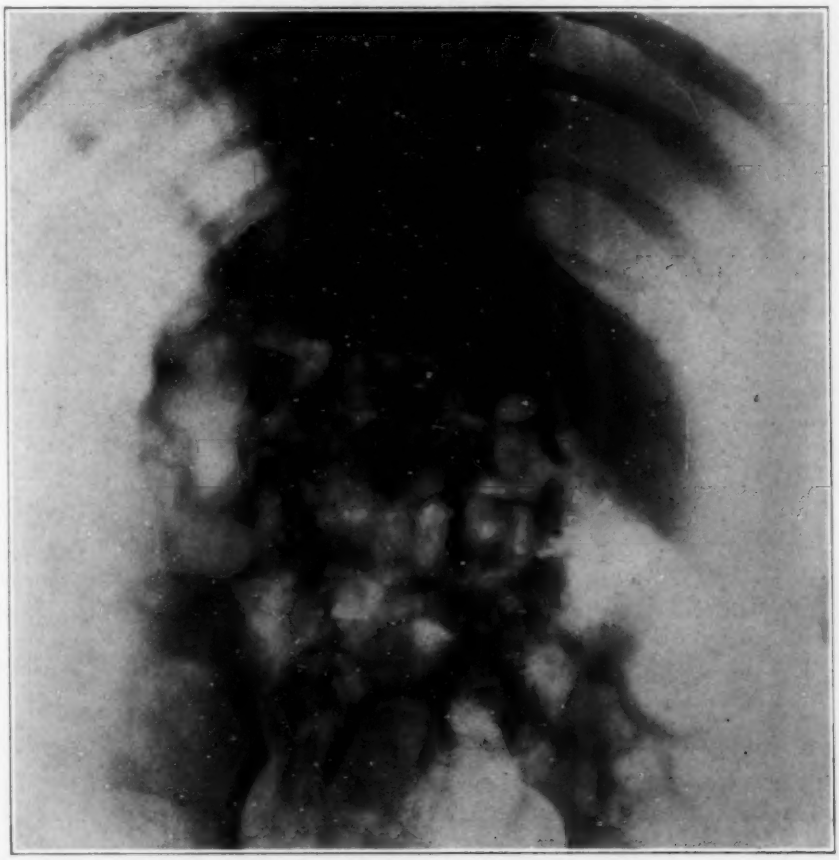

Fig. 7.—Roentgenogram of same spine as in Figure 6 taken eighteen months later. Note growth of the defect, also fusion of the ninth and tenth ribs.

CASE 13.—Death occurred on the ninth day after operation from meningitis.

CASE 14.—Death occurred on the eleventh day after operation from hydrocephalus and infection.

CASE 15.—Death occurred with fever, cyanosis, rapid respirations and weak pulse. There was atrophic ulcer on the right knee.

CASE 21.—Death occurred twelve days after operation. The temperature was subnormal, and the cause unknown.

Case 24.—Death occurred on the day after operation from acute hydrocephalus; the temperature rose to 107 F.

Case 33.—Shortly after operation the fontanels bulged, the patient had a convulsion with retraction of the neck, and died three days later from hydrocephalus, and possibly meningitis.

Case 34.—Death occurred seventeen days after operation, from hydrocephalus.

Case 36.—Death occurred thirty-three days after operation from bronchopneumonia, and slight hydrocephalus.

Fig. 8 (Case 10).—Appearance of patient at time roentgenogram (Fig. 7) was taken. She died of meningitis at the age of 19 months.

Case 38.—Death occurred on the third day after operation from loss of cerebrospinal fluid and possibly meningitis.

Case 39.—Death occurred twelve days after operation from meningitis and septicemia.

Case 2.—In this case the result was unknown.

COMMENT

Of the thirty-nine patients operated on, twenty-two were discharged, sixteen died, and in one instance the result was not known. This gives an operative mortality of 41.02 per cent. If we include the unknown

result as a possible death, the mortality would be 43.58 per cent. The causes of death were: six, hydrocephalus; three, hydrocephalus plus meningitis; two, meningitis; two, leakage of cerebrospinal fluid; two, cause unknown (one a few hours after operation, cause not determined, one on the twelfth day after operation, with subnormal temperature) and one brochopneumonia on the thirty-third day after operaion.

FOLLOW UP RECORD AND END RESULTS

CASE 3.—The patient was seen in the admitting room two months after operation. There was purulent discharge from the wound. No reply was received to a letter sent on October, 1923. (Result unknown—probably died.)

CASE 5.—The patient developed some hydrocephalus, which became arrested spontaneously. June 16, 1922 (four years after operation) the patient was paralyzed and had trophic ulcers; there was no bulge at the back. (Living, but had complications.)

CASE 6.—The patient was discharged from the hospital relieved; he had progressive hydrocephalus and died May 28, 1919, nine months after operation.

CASE 9.—The patient was discharged well. A letter received, Oct. 26, 1923 (five and one twelfth years after operation), stated: "The child runs and plays normally. There is hair present at the site of the scar."

CASE 12.—The patient was discharged from the hospital relieved. According to on outpatient note, there was slight hydrocephalus, which became arrested spontaneously. A letter received three and ten twelfths years after operation, stated: "Child getting along pretty well."

CASE 16.—The patient was discharged well. On March 15, 1921, four months after operation, the child was well. A letter received Oct. 29, 1923, three years after operation, stated: "Child in the best of health."

CASE 17.—The patient was discharged relieved. Feb. 17, 1921, one month after operation, there were increased signs of cerebrospinal fluid discharge; convulsions; no vomiting; increase in the size of the head in all directions. No reply was received to letters.

CASE 18.—The patient was discharged relieved, in good condition. No reply was received to letters.

CASE 19.—The patient was discharged well. A letter received in October, 1923, two and seven twelfths years after operation stated: "Patient the picture of health."

CASE 20.—The patient was discharged relieved. A letter received on Nov. 10, 1923, stated: "General condition good, walks all right but if he falls cannot get up, never creeps. When he grips with the right hand the left works also. He is mentally bright."

CASE 22.—The patient was discharged relieved. Oct. 27, 1921, one month after operation there was improvement in power of the legs; slight intracranial pressure; Jan. 10, 1922, four months after operation, slight hydrocephalus, general condition good. No reply was received to letters.

CASE 23.—The patient was discharged well. A letter in October, 1923, stated: "Patient died from pneumonia."

CASE 25.—The patient was discharged well. No reply was received to letter of October, 1923. (Unknown.)

CASE 26.—The patient was discharged relieved. An outpatient note stated that the general condition was excellent. A letter received, October, 1923 (one and six twelfths years after operation), stated: "Since discharge from the hospital my baby has improved wonderfully."

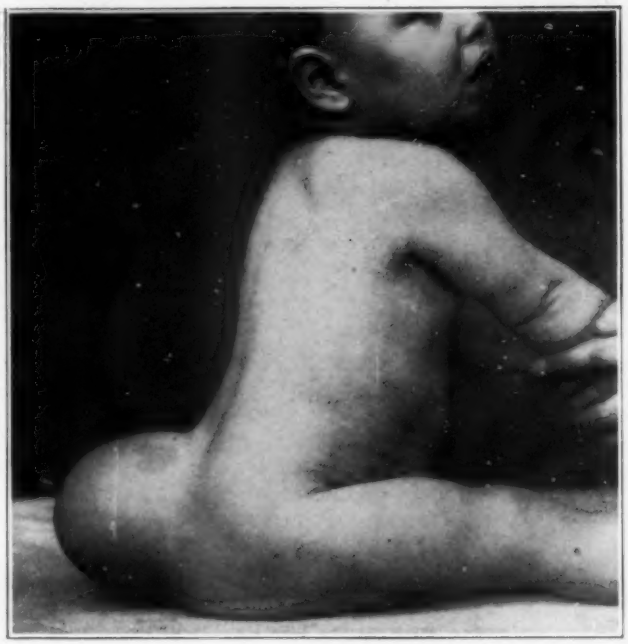

Fig. 9.—Operation series, Case 37, illustrating a low spina bifida. This was a sacral meningomyelocele with some paralysis.

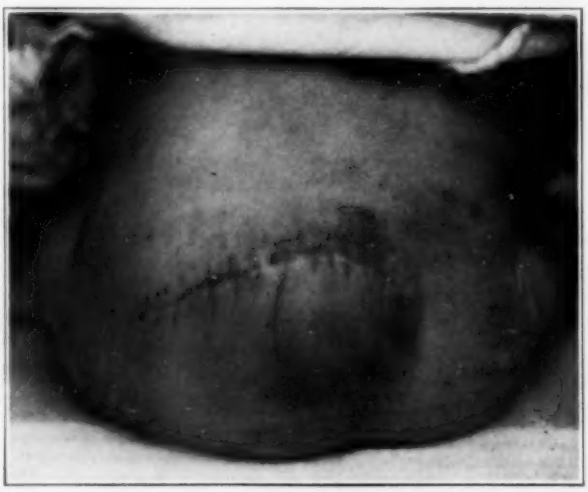

Fig. 10.—Postoperative result, Case 37. Note horizontal closure of skin.

CASE 27.—The patient was discharged relieved. A letter received Nov. 1, 1923, one and six twelfths years after operation, stated: "Legs paralyzed; incontinent. Child doesn't talk much yet."

CASE 28.—The patient was discharged relieved. June 24, 1922, wound had healed, and hydrocephalus was increasing. A letter received in October, 1923, stated that the patient was incontinent of feces; the head was large; the spina bifida had healed.

CASE 29.—The patient was discharged relieved, with slight bulging of the fontanel. A letter in October, 1923, stated: "Patient died in convulsions two weeks after leaving the hospital."

CASE 30.—The patient was discharged relieved to the orthopedic service (for associated Pott's disease). A letter in October, 1923, stated: "Patient died from pneumonia."

CASE 31.—The patient was discharged relieved. A letter in October, 1923, stated: "Seems normal in every way."

CASE 32.—The patient was discharged relieved. A letter in October, 1923, stated: "Child now is 3 years and 2 months old, and is perfectly well."

CASE 35.—The patient was discharged relieved. No reply was received to letters.

CASE 37.—The patient was discharged relieved. Dec. 15, 1923, there was some bulging and some paralysis.

END-RESULTS

A follow-up was carried out on the twenty-two patients who had been discharged from the hospital living. Twenty-four letters were sent out; thirteen replies were received, and one letter was returned unclaimed, the family having moved away. In six cases, the end-result is unknown, and the end-results of the remaining four were taken from the last notes in the outpatient department record.

Unknown	6
Living and well	7
Living and improving	1
Living with complications	4
Dead	4
Total	22

INOPERABLE CASES

The ages of patients in this group when first seen ranged from 12 hours to 1½ years. There were thirteen girls and five boys. Sixteen were meningomyeloceles and two meningoceles; all were located in the lumbar region with the exception of one dorsal meningomyelocele, one cervical meningocele, one dorsolumbar meningomyelocele and one lumbosacral meningomyelocele.

Fifteen patients showed definite paralysis; in three, it was not observed (two meningoceles, one cervical and one lumbar and one lower thoracic—the last patient was operated on at the Boston City Hospital and died shortly after). Seven patients showed hydrocephalus. All but two cases were considered inoperable; one of the patients had

a small lumbar meningocele that was doing well enough at the time without operation; the other had a cervical meningocele and was recommended for operation, but two weeks after the visit to the outpatient

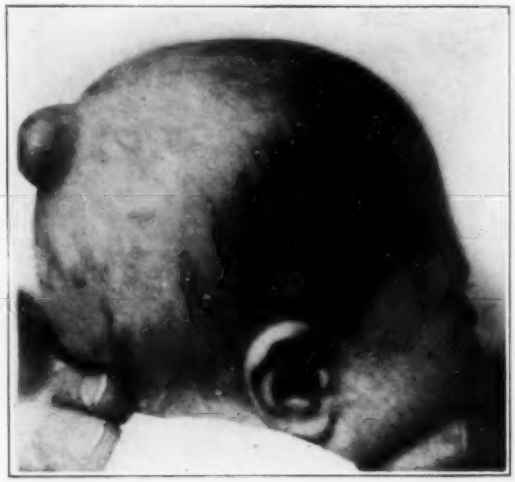

Fig. 11.—Cephalocele series, Case 4, excision with recovery.

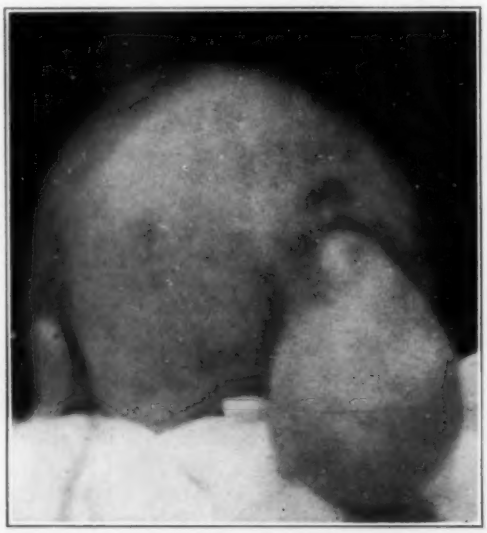

Fig. 12.—Cephalocele series, Case 5, excision followed by hydrocephalus.

department the child became ill, had convulsions and died on the way to the hospital.

In one case a maternal aunt had spina bifida.

FOLLOW-UP AND END RESULTS

CASE 1.—The last outpatient department note when the child was 16 days old stated: "Sac ruptured, temperature 102 F. No reply to letter."

CASE 2.—The patient was referred to the social service for care. A letter in October, 1923, stated: "Admitted to the New England Hospital. Lived sixty-six days and died of meningitis."

CASE 3.—The sac was mistaken and opened for an abscess shortly after birth. The child, aged 2 months, was admitted to the hospital at 10 a. m. and died at 12:30 p. m.

CASE 4.—The patient was seen in the outpatient department with a small lumbar meningocele. In April, 1923, aged 3 5/12 years, there was no change and there were no symptoms. A letter in October, 1923, stated the child was well.

CASE 5.—No reply was received to letters.

CASE 6.—The child was seen again at the age of 6 weeks in the outpatient department. Convulsions occurred at times. The wall of the sack was thicker. A letter in October, 1923, stated: "Died at the age of 2 months, cause unknown."

CASE 7.—No reply was received to letter.

CASE 8.—A letter in October, 1923 (age of patient 1 9/12 years), stated: "General condition very good. Hydrocephalus not progressing. Spina bifida sac healed."

CASE 9.—A letter, Nov. 12, 1923, stated: "Lived six months, sac ruptured three times and the last time filled up to the height of 4 inches. Patient lived one week after it ruptured the last time, and died from loss of cerebrospinal fluid and meningitis."

CASE 10.—The patient was first seen, July 27, 1921, with a large raised meningomyelocele, and was admitted to the hospital, March 29, 1923. The temperature was 106 F., and there were rigidity of the neck and positive Kernig sign. A diagnosis was made of meningitis. The patient was discharged, April 23, 1923. A letter, Jan. 8, 1924, stated: "Patient died one week after leaving hospital at the age of 9 9/12 years."

CASE 11.—The patient was first seen, May 8, 1923, with a lower thoracic meningomyelocele discharging cerebrospinal fluid, and hydrocephalus. Seen Aug. 29, 1923. The hydrocephalus was arrested, the spina bifida sac large. A letter in January, 1924, stated: "Operation at Boston City Hospital at 6 months and patient died shortly afterwards."

CASE 12.—The patient was first seen, June 12, 1923, with a redunculated cervical meningocele having a glioma on the summit, and was recommended for operation. A letter, Oct. 31, 1923, stated: "Two weeks after visit to outpatient department became ill and died on the way to the hospital in convulsions."

CASE 13.—The patient was first seen, June 12, 1923, at age of 1 7/12 years, with a large meningomyelocele which was increasing in size, and discharged cerebrospinal fluid at times. The patient had paralysis and hydrocephalus. A letter in October, 1923, stated: "Lived to the age of 1½ years and died from loss of cerebrospinal fluid and meningitis."

CASE 14.—The patient was first seen, July 31, 1923, at the age of 3 months, with a large meningomyelocele discharging cerebrospinal fluid. Paralysis and hydrocephalus were also present. A letter Oct. 30, 1923, stated: "Died one week after visit to the hospital from loss of cerebrospinal fluid."

CASE 15.—First seen Oct. 28, 1923, aged 1 day, with large sessile lumbar meningomyelocele, unruptured, transparent sac. Paralysis was present. The patient lived only twenty-four days.

Case 16.—The patient was first seen, Jan. 23, 1923, aged 13 days, with a large oval, sessile dorsolumbar meningomyelocele with a thin infected sac. Paralysis and hydrocephalus were present. The case was referred to the social service visiting nurse.

Case 17.—The patient was first seen, June 17, 1923, at the age of 1 day, with a lumbar meningomyelocele, thin skinned, unruptured and raised. Paralysis was present but no hydrocephalus. He was discharged from the hospital, without operation, June 21, 1923. Seen in outpatient department, Nov. 9, 1923, he had convulsions and strabismus with hydrocephalus of slight degree. January, 1924, he was doing fairly well; the sac was large.

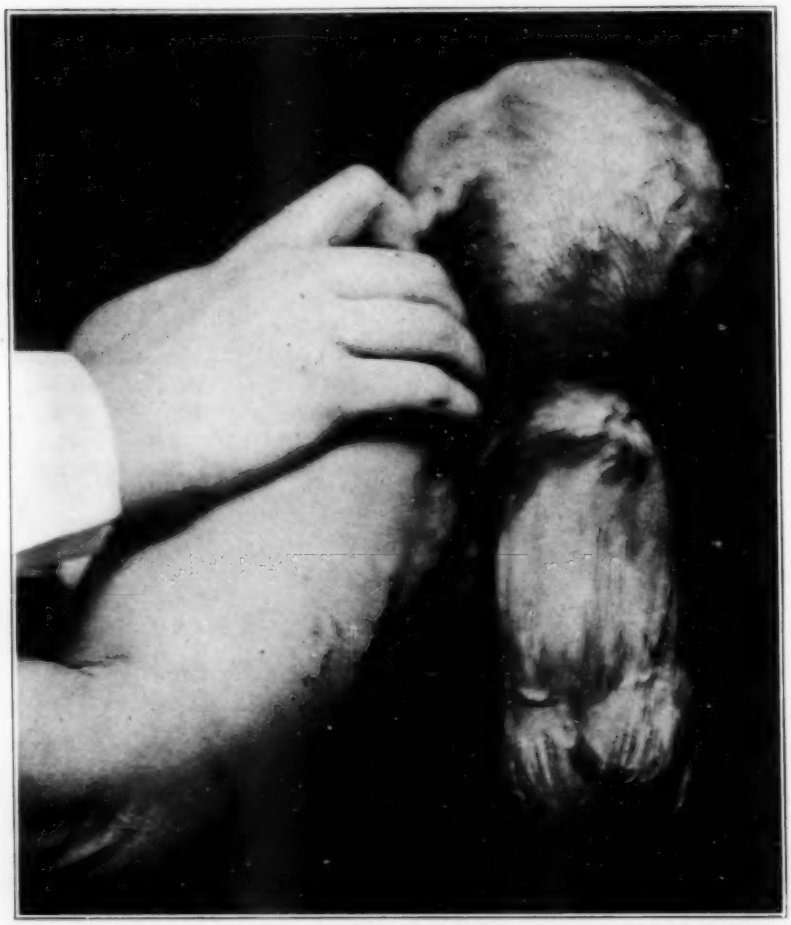

Fig. 13.—Cephalocele series, Case 1; parents refused operation; end result unknown.

Case 18.—The patient was seen, Dec. 4, 1921, at the age of 2 days, with a large lumbosacral, thin-walled, sessile meningomyelocele—three arches defective. Ulceration was present, and there was slight paralysis. No operation was

advised. The patient was seen, Feb. 11, 1922, when hydrocephalus was developing. No operation advised. Later the child was taken into the Boston City Hospital. The end result is unknown.

Dead	10
Living	2
Possibly living	2
Unknown (probably dead)	4
Total	18

CEPHALOCELES

One patient had meningo-encephalocele; the other four presented cranial meningoceles of various sizes. They were all located in the region of the posterior fontanel. The four meningoceles were treated by operation, and the meningo-encephalocele patient was taken from the hospital untreated at the parents' request. Of the four patients operated on, one died of hydrocephalus, two are well, the fourth has survived operation ten days but is developing hydrocephalus fairly rapidly. The subsequent history of the meningo-encephalocele is unknown.

CONCLUSIONS

1. Fifty-seven patients with spina bifida and five patients with cephalocele afforded the material for this study.

2. The high mortality rate of spina bifida and the futility of attempting to cure the condition in certain cases by radical operation is again emphasized.

3. Removal of the spina bifida sac and closure of the defect probably attacks only the effect and not the cause of the condition.

4. We must recognize in addition to a faulty anatomic relation of tissues the rôle which the cerebrospinal fluid mechanism undoubtedly plays in the production and behavior of the spina bifida. Clinical evidence demonstrates the safety valve action of the sac.

5. Early radical operation may prevent deaths from infection or leakage of cerebrospinal fluid, but the frequent incidence of hydrocephalus and death after removal of the sac is cause for a guarded prognosis.

6. The patients treated in this series were all under 3 years of age. Keen has pointed out the fact that "the mortality is higher in operations done in the first few months, 35 + per cent., against 4.7 per cent. in those 5 years old and over. This simply means that, by waiting, natural causes have produced the mortality, part of which might have been attributed to surgery, and the latter has lost the opportunity of curing or improving some cases."

7. The contraindications to operating are: hydrocephalus, extensive paralysis, and infected sacs.

8. The mortality in the thirty-nine patients personally treated by operation was 43.58 per cent. Hydrocephalus, meningitis and leakage of cerebrospinal fluid were responsible for the postoperative deaths.

9. Following up the twenty-two patients discharged from the hospital living, it was found that seven are living and well, one living and improved, four living and have complications, six are unknown, and four are dead (two died of penumonia).

10. Of the eighteen patients whose condition was considered inoperable, ten died, two are known to be living and of the six whose condition is unknown, two are possibly living, and four are probably dead.

11. Small cephaloceles with thin membranes should be operated on to prevent infection and leakage. There is always danger of hydrocephalus resulting after operation.

12. The patients in this series have not been followed long enough to develop all the complications of this distressing condition. If those who are paralyzed and incontinent live long enough, they are usually afflicted with cellulitis, sinuses about the buttocks and trophic ulcers. If fatal intercurrent disease does not supervene, they finally die of urinary tract infection.

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DISCUSSION

DR. PERCIVAL BAILEY: Dr. Cutler has admitted that there exists in these children a defect of the normal mechanism for absorption ofthe cerebrospinal fluid and that he considers the meningocele as a safety valve for the absorption of fluid. From this point of view his operative procedure seems scarcely rational, for it consists essentially in sewing up the safety valve. The result is just what one would expect—the development of a hydrocephalus.

All operative procedures should be based on a thorough understanding of the pathologic alteration that one seeks to correct. Certainly the mechanism of production of meningocele and its associated hydrocephalus are by no means understood, and a thorough investigation is badly needed.

In the present state of our knowledge, it would seem that one should attempt to deal first with the hydrocephalus when it can be demonstrated, and when the condition of the meningocele makes closure imperative to prevent rupture and infection, then the hydrocephalus must be dealt with secondarily as soon as it makes its appearance.

A DEVELOPMENTAL ANOMALY OF THE CEREBRAL CORTEX *

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AND

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We report the following case because of the evident rarity of occurrence of this developmental anomaly.

REPORT OF CASE

History.—E. A., a man, aged 53, was admitted to St. Michael's Hospital, Toronto, in October, 1923. He was a diabetic and was receiving insulin treatment. On admission he was acutely ill with pneumonia, to which he succumbed within two days after entering the hospital. There was nothing in his medical history to suggest mental defect or disease.

Necropsy Findings.—There were the scars of old tuberculous disease at the apex of the right lung. This had evidently "lit up," because the bronchopneumonia, of which he died, was proved to be tuberculous. No other pathologic lesion was found in the chest or abdomen.

The skull was opened, and the brain removed. Coronal sections of the frontal lobes disclosed the presence of multiple gray masses scattered irregularly through the white matter of the left frontal lobe. The right frontal lobe was normal, and there was no edema of the brain substance. The anterior horns of the lateral ventricles appeared normal and equal in size, and the cerebrospinal fluid contained in them showed no differences between the right and the left sides either in appearance, amount or tension. The brain showed no other defect or disease. The cerebral arteries were normal.

After fixation and hardening in formaldehyd, a more detailed examination was instituted. This consisted of a macroscopic study of a series of vertical transverse sections, a photograph of one of which is reproduced in Figure 1. For microscopic examination paraffin and celloidin blocks of the gray areas were prepared. The latter were used for the Weigert-Pal technic. Sections of the former were stained as follows: (*a*) with methylene blue for the cellular content of the anomalous masses; (*b*) with Mallory's triple stain (orange G, acid fuchsin and aniline blue) for the connective tissue element; (*c*) with Mallory's phosphotungstic stain for neuroglia, and (*d*) with hematoxylin and eosin.

Macroscopic Appearances.—Figure 1 shows the posterior surface of a vertical transverse section of the hemispheres cut through the anterior horns of the lateral ventricles. The white matter of the left frontal lobe is seen to contain irregular dark masses which have the appearance of gray matter. The inner-

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most of these masses raise up the ependyma of the ventricular cavity so as to give its lateral wall a tubercular contour. The cavity of the anterior horn seems somewhat larger on the left, but it is doubtful whether any definite internal hydrocephalus was present. The left frontal lobe is somewhat smaller than the right, but this was not sufficiently noticeable to cause any remark at necropsy.

It will be noted that the corona radiata of the right frontal lobe is completely free from these anomalous deposits.

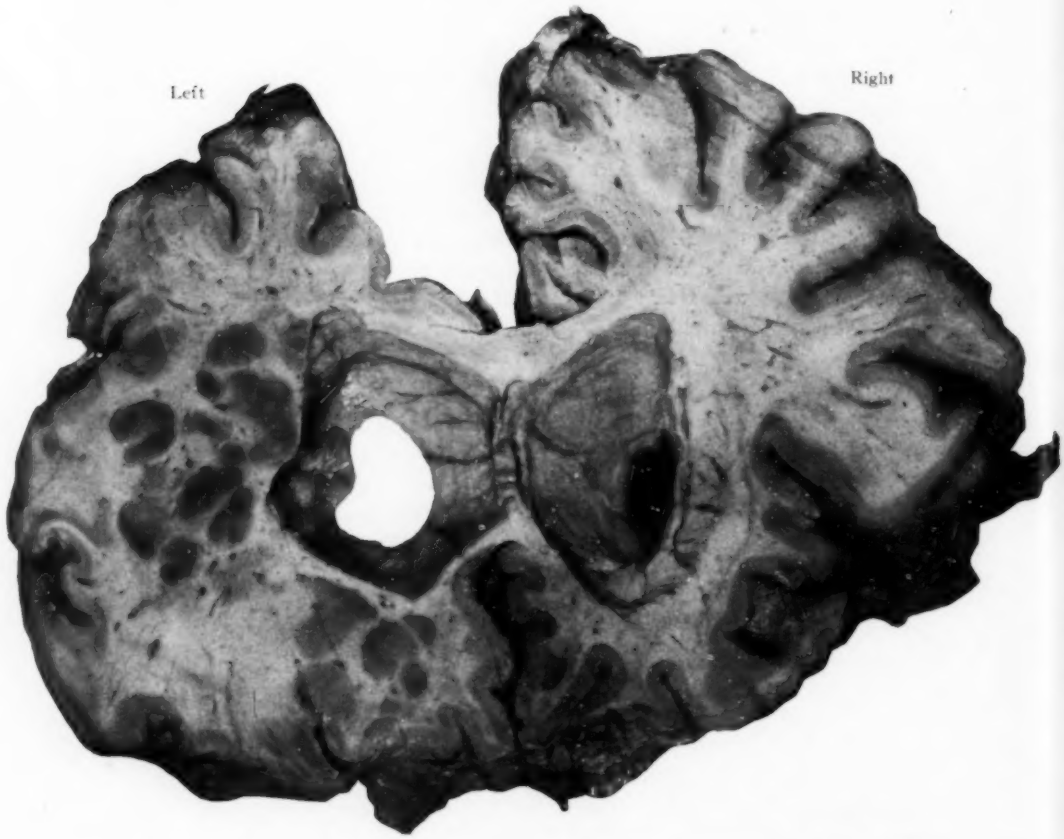

Fig. 1.—Posterior surface of vertical transverse section of hemispheres. Anomalous gray deposits shown scattered through left frontal white matter. Subependymal masses protrude into the cavity of the left lateral ventricle interrupting the normal smooth contour.

The whole of the left frontal and parietal lobes of white matter was permeated by the masses. There was nothing in any of the sections to suggest that they owed their origin either to the cortex cerebri or to the caudate nucleus.

Microscopic Examination.—A section of one of the gray masses showed the presence of nerve cells, varying considerably in type, lying in a matrix of delicate fibers. The fibers did not give a positive reaction with the neuroglia stain employed, and so were probably not neuroglia cell processes (Fig. 2).

The cell content, as examined with an oil-immersion lens in sections stained with methylene blue, showed cells of three fairly distinct types which seemed to mark three stages in the development of a neuroblast (Fig. 3).

The first type showed a large round nucleus containing well marked chromatin granules and with extremely little surrounding protoplasm.

In the next stage, the nucleus stained more deeply blue, tending to obscure the granules, and the cytoplasm was greater in amount.

The third type of cell was definitely pyramidal in shape, with a deeply staining spherical nucleus and abundant cytoplasm.

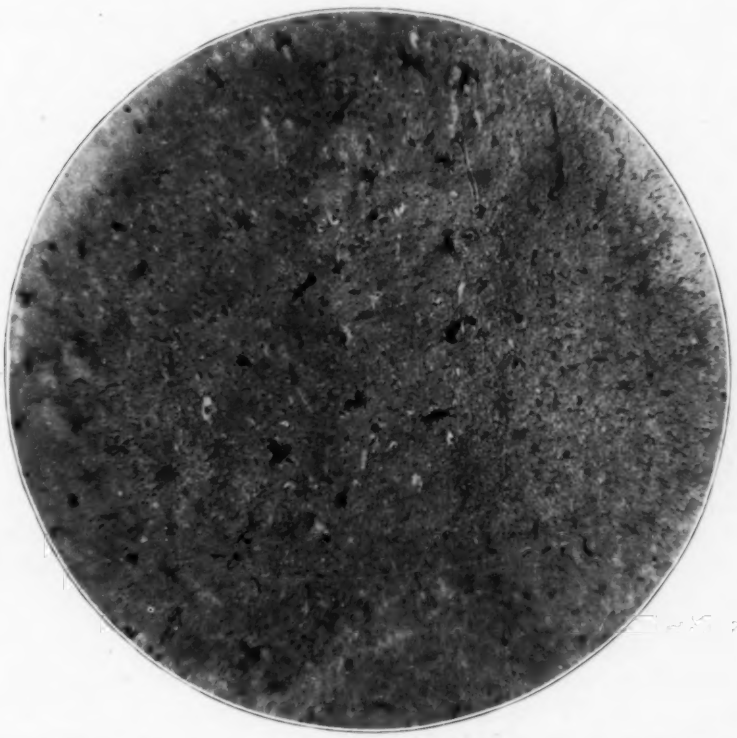

Fig. 2.—Low power view of a gray area, showing a circumscribed area in the center of the field filled with nerve cells surrounded by normal white matter. The larger black areas are masses of stain deposit. × 94; Zeiss; Mallory's phosphotungstic stain.

Nissl granules were not to be seen even in the largest pyramidal cells of these areas, from which it may be inferred that none of these nerve cells was completely developed.

Although the gray areas showed sharply delimiting edges to naked eye examination, this was not altogether borne out by microscopic observation. Isolated nerve cells of the third or pyramidal type mentioned above could be found lying in the normal white matter separating the gray areas. A gray mass which showed a well marked edge in one direction faded off into the white matter in another, the masses of cells being separated by normal white fibers.

DISCUSSION

Immediately after its closure, the neural tube shows three histologically distinct areas which, followed from within outward, are known as the ependymal, mantle and marginal zones. The mantle zone contains the primitive nerve cells or neuroblasts and is separated from the central cavity of the tube by only a single layer of ependymal cells. The mantle zone as development proceeds becomes thickened on the lateral aspects of the tube and soon shows a differentiation into a basal or ventral and an alar or dorsal lamina. Early in fetal life the

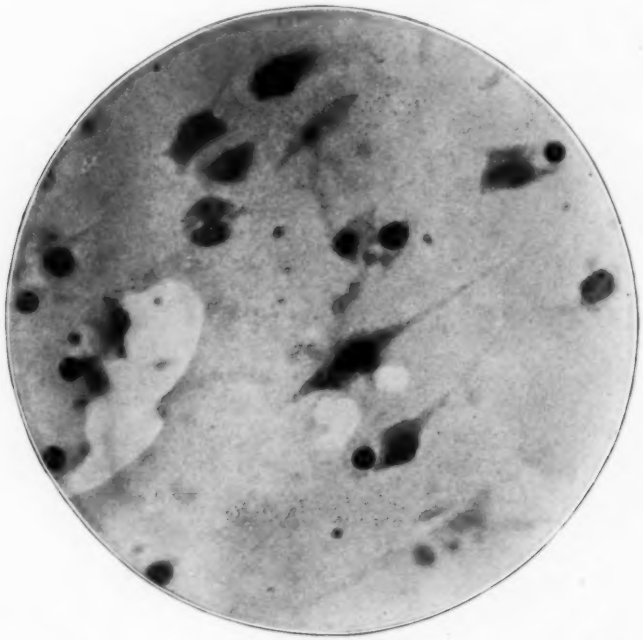

Fig. 3.—High-power view of an anomalous gray mass, showing the three stages of development of a neuroblast. × 1,100; Zeiss; methylene blue stain.

head end of the neural tube shows three expansions of its cavity to form the three primitive cerebral vesicles. In the third month of intrauterine life, the alar lamina of the mantle layer of the foremost part of the forebrain vesicle shows a change in its histologic arrangement. This change is the migration outward of its neuroblasts through the marginal zone of the tube wall until they come to lie as layers of undifferentiated nerve cells immediately under the surface of the primitive prosencephalon. The migration of cells to the surface is completed by the end of the fourth month, and thus is accomplished the first stage in development of the cerebral cortex.

The subsequent changes which take place to form the normal adult cortex from these undifferentiated layers of neuroblasts are assumed by Vogt to be: first, a grouping of the elements, and second, a ripening of the elements. These two changes he calls the organogenetic development of the cortex. The neuroblast elements first become grouped into tangential layers of similar cells and, when thus layered, ripen into their adult form.

It would appear, therefore, that in the case under consideration some abnormal process acting during the fourth month of fetal life had, to a limited extent, checked the outward migration of neuroblasts in the left side of the forebrain vesicle. Although there was no trace of grouping or layering of these stranded groups of elements, some attempt at ripening was seen in the formation of immature pyramidal cells.

Von Monakow,[1] in 1899, and Vogt,[2] in 1905, published the results of their studies of cases of displaced cerebral cortex. They both gave a classification of the observed types of this anomaly, and these classifications are so similar in their essential features that they may be fairly combined in the following list of types:

I. Malposition of cell individuals. The cell found varies from a small granular cell to a small pyramid.

II. Closed cell-groups. Heaps of undifferentiated nerve cells scattered in the white matter.

III. Clumps of gray matter under the ependyma. These, von Monakow states, show neuroglia cells, neuroblasts and occasionally mature ganglion cells.

IV. Dislocated areas showing cortical layering.

V. Areas of normally placed cortex showing abnormal or imperfect layering.

VI. Paradoxical architecture. Von Monakow adds this group to include cases in which he has found the cell content normal but the order of the layers displaced. Occasionally an apparently normal piece of cortex has been found placed obliquely to the surface of the hemisphere.

The first three types of anomaly are illustrated in the case under discussion.

Oseki,[3] in 1913, reviewed the literature on this subject and described three personal cases: (1) the case of a new-born child, of which there

1. Von Monakow, C.: Ergebnisse der allgemeinen Pathol. u. pathol. Anat., 1899.

2. Vogt, T.: Arbeiten aus dem Hirnatomischen Institut in Zurich, Part 1, 1905.

3. Oseki, S.: Beitrag zur Heterotopie der grauen Substanz in Gehirn, Monatsschr. f. Psychiat. u. Neurol. **34**, 1914.

are no clinical particulars; (2) the case of a mentally normal woman, aged 75, in whom necropsy revealed besides the anomalous cortex, diffuse bronchitis, nephritis, obsolete tuberculosis of the lungs and peribronchial glands, and arteriosclerosis; (3) the case of a female idiot who died at the age of 2 of enteritis. Postmortem examination revealed hydrocephalus. In each of these brains the gray deposit was subependymal in position. It was situated in the anterior horn and body of the left ventricle in Case 2 and in both anterior horns in Case 3. The exact site is not mentioned in the first case.

Microscopically he failed to find pyramidal cells in Case 1, and, in regard to the cortex overlying the deposit, he states that the first layer was fibrous and that, below the second layer, the cortical structure was indefinite.

In Cases 2 and 3, he describes typical pyramidal cells, glia cells and glia fibers. Medullated nerve fibers were found running through the masses. He says that the convolutions were normal in his third case.

It would seem, therefore, that whereas the infant showed a picture typical of arrest of cortical development at the fourth month, the second and third patients displayed some attempt at ripening of the nerve elements in their abnormal situation.

Oseki's second case is of interest in that the woman is described as mentally normal, which is not usual, although there was no history of mental defect in our case. In the great majority of case reports in the literature, mental disease is recorded. Diagnoses of idiocy, epilepsy, melancholia, mental deficiency and dementia are the rule.

Ermann,[4] however, describes a mentally healthy woman, aged 62, who had gray deposits in the floor of a lateral ventricle.

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4. Ermann: Heterotopie grauen Hirnsubstanz, Virchows Arch. f. path. Anat. **56**:419, 1872.

LOCULATED MENINGITIS

WITH THE SYNDROME OF FROIN IN THE SPINAL FLUID *

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The occurrence of fibrinogen in the spinal fluid of patients with acute meningitis was observed toward the end of the last century. In 1903, Lépine [1] noted the formation of a coagulum in spinal fluid from a patient whose case was diagnosed as spinal rheumatism. During the same year Froin [2] reported three cases in which he had observed the spinal fluid changes which as a syndrome have since borne his name. The syndrome of Froin consists of a yellowish or xanthochromatic spinal fluid with increased globulin and albumin, and showing the property of spontaneous coagulation. The coagulum usually forms within one-half hour after the withdrawal of the fluid. Frequently a pleocytosis is present in the fluid. In 1910, Nonne [3] called attention to the increased globulin occurring in the spinal fluid of patients with spinal cord tumors. Many variations of these pathologic conditions are found in spinal fluids. The French have devoted more attention to the yellowish discoloration and massive coagulation, while the Germans have placed most emphasis on the increased globulin and albumin.

In Froin's [2] original paper, he did not attempt to draw conclusions from his cases. In 1904, Cestan and Ravaut [4] reported a case of flaccid paraplegia with spinal fluid changes of this type. This case at necropsy showed a meningomyelitis involving the sacral enlargement of the cord; also a fibrinous concrescence of the meninges and cord at the level of the conus, due to pachymeningitis. They considered the cause of the spinal fluid syndrome to be meningeal inflammation and associated hemorrhage. This explanation of Cestan and Ravaut [4] was rejected by Sicard and Descomps.[5] These authors found that fluid obtained by a second lumbar puncture, performed a few days after the first one showed a much lower albumin content. They called particular attention to the changeableness of the fluid findings at different punctures. Necropsy in their case revealed a mass of fibrous and caseous tissue in the epidural space at the level of the eleventh and twelfth thoracic

* From the Department of Internal Medicine, University of Michigan Medical School, Ann Arbor.

1. Lépine, J.: Lyon méd. **101**:298-302, 1903.
2. Froin, G.: Gaz. d. hôp. **76**:1005, 1903.
3. Nonne, M.: Deutsch. Ztschr. f. Nervenh. **40**:161-167, 1910.
4. Cestan, R., and Ravaut, P.: Gaz. d. hôp. **77**:985-988, 1904.
5. Sicard, J., and Descomps, P.: Gaz. d. hôp. **81**:1431, 1908.

vertebrae. The dura was adherent to the arachnoid in several places. They considered the inflammation to be annular in form, thereby causing vascular compression. Sicard and Descomps first recognized the significance of a closed-off or loculated meningeal pouch as a cause of the xanthochromia and coagulation. The vascular compression and local edema were, to their minds, but minor elements in the causation of these unusual changes in the spinal fluid. They considered the plasma to be an exudate into the loculated space in which the plasma and corpuscular elements were retained. They thought the variation in the fluids drawn at intervals was due to the filtering through of the normal fluid from above into the closed pouch below.

Few necropsies have been performed on patients who, previous to death, have shown these spinal fluid changes. However, nearly all cases which have come to necropsy have shown this loculation of the meninges with the separation of the spinal canal into an upper and a lower segment.

The syndrome of massive coagulation with xanthochromia has been observed in a great variety of conditions. It was found in patients having a condition resembling Landry's paralysis by J. Donath [6] and Renon and Monier-Vimard.[7] Claud [8] noted these changes in the spinal fluid of a patient suffering from alcoholic polyneuritis. The syndrome of Froin has frequently been found in the spinal fluid in Pott's disease, Sicard, Foix, and Salin,[9] having been the first to note this association. In 1910, Raven [10] collected 145 cases from the literature in which the presence of Froin's syndrome in the spinal fluid was due to compression of the cord by: (1) a tumor, (2) diseases of the vertebrae, or (3) pachymeningitis. The occurrence of these spinal fluid changes in epidemic cerebrospinal meningitis was observed by Forbes and Adam [11] in 1915. Since then numerous similar examples have been reported, especially in the French literature. The direct cause of the meningitis, however, is often obscure. In a case reported by Greenfield,[12] with xanthochromia and massive coagulation in the spinal fluid, the etiologic agent appeared to be a staphylococcus. Frequently these spinal changes have been considered to be due to tuberculosis, although this has never been proved. More frequently the condition has occurred in patients

6. Donath, J.: Wien. klin. Wchnschr. **18**:1327-1330, 1905.

7. Rénon, L., and Monier-Vimard, R.: Compt. rend. Soc. méd. d. hôp. de Paris **28**:32-39, 1909.

8. Claud, H.: Rev. neurol., Paris **19**:1420-1422, 1909.

9. Sicard, J.; Foix and Salin, H.: Presse méd. **18**:977, 1910.

10. Raven, W.: Deutsch. Ztschr. f. Nervenh. **67**:55, 1920.

11. Forbes, D., and Adam, D. C.: Pub. Health, London **28**:225-235, 1914-1915.

12. Greenfield, J. G.: J. Neurol. & Psychopath., Bristol **2-7**, 1921.

with syphilis. In a case reported by Babinski,[13] definite improvement occurred under antisyphilitic treatment.

The French literature on this subject has been comprehensively reviewed by Mix.[14] The subject of Froin's syndrome has been brought up to a recent date by the reviews of Greenfield [12] and Lantuéjoul.[15] In Lantuéjoul's review, he finds only thirty-five cases in the literature which strictly conform to the full syndrome, excluding all those cases in which the fluid did not coagulate massively and spontaneously. In the present report, I have considered only those cases in which the diagnosis of syphilis was probable or proved. I have considered this type of case only as suitable for comparison with the case which I wish to report.

REPORT OF CASES

Case 1 (Froin's [2]).—A woman, aged 36, had right facial paralysis, which occurred in November, 1902. In April, 1903, a painful flaccid paraplegia occurred. Four lumbar punctures were performed on this patient. During the first puncture, the fluid ran slowly into the tube; it was clear and coagulated spontaneously into a solid mass. After the clot had retracted, the centrifuged specimen showed a lymphocytosis. The fluid withdrawn at the second puncture was slightly yellowish, coagulated spontaneously and contained increased lymphocytes. The third fluid was clear. Guinea-pigs inoculated with this fluid for the presence of tubercle bacilli were found to be negative. Fluid withdrawn at the fourth puncture was slightly yellowish and coagulated within one hour after withdrawal. The cerebrospinal fluid was never under notable tension; it varied in its color; it always contained increased lymphocytes and coagulated spontaneously, or showed a coagulum within one hour. This case apparently had a pachymeningomyelitis which involved the conus terminalis and was due to syphilis.

Case 2 (Babinski [13]).—This patient showed a spastic paraplegia. Because of the presence of contractures, exaggerated reflexes, Babinski toe sign and the sphincter disturbances, the diagnosis of pachymeningitis was considered probable. The sensations were only slightly disturbed. The spinal fluid after withdrawal showed the characteristics of Froin's syndrome. This patient was treated by mercuric inunctions, with slight improvement. There was some improvement noted following the lumbar puncture with the withdrawal of the spinal fluid. The case was diagnosed as a fibrinous and hemorrhagic pachymeningitis, probably due to syphilis.

Case 3 (Froin [16]).—This patient showed multiple lesions of the spinal cord, which were probably syphilitic. There was optic atrophy with paralysis of the external rectus muscle, and Argyll Robertson pupils. The symptoms in the legs were pronounced, the severe pains occurring first in the left and later in the right leg. Subsequently, there was atrophy of the left leg with a loss of the spinal reflexes, and a slight Kernig's sign. The spinal fluid showed the changes typical of Froin's syndrome, with lymphocytosis.

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13. Babinski, J.: Guérison, Soc. méd. d. hôp. **20**:1083-1089, 1903.
14. Mix, C. L.: Surg. Clin. of J. B. Murphy **4**:317-382, 1915.
15. Lantuéjoul, P.: Rev. neurol. **36**:339, 1920.
16. Froin, G., quoted by Aubrey: Thèse de Paris, 1909.

Case 4 (Klieneberger[17]).—A man, aged 26, had had severe pains in his right leg, with a dragging of the right foot, since the age of 19. These conditions were improved by massage and electricity. He was married at the age of 24, and during the same year had typhoid fever, followed by disturbance in emptying his bladder. During the convalescence following the typhoid fever, he first noticed clumsiness in using his left hand, with the occurrence of slight pain in the same extremity. The right leg became definitely weaker than the left, and the left arm and right leg showed slight atrophy. Two years later he began to have weakness and pains in his neck with cramps in the right leg. Within a few months after the onset of the latter symptoms, pains occurred in the right leg associated with weakness. Within a short time he had radiating pain occurring in his back, followed shortly by spasm of his bladder sphincter, which necessitated catheterization. Examination of the patient revealed sensitiveness over the sixth to the eighth dorsal vertebrae, and ankle clonus in both extremities, but mostly in the left. There was a bilateral Babinski sign and absence of the abdominal reflexes, the cremasteric being present. The spine was very stiff, and there was slight atrophy in the muscles of the left hand associated with awkwardness in its use. The right leg was dragged. Passive movements of the extremities were not interfered with, but there was ataxia in both legs. The tendon reflexes in the arms were increased. There was hyperalgesia in the right arm and hypalgesia in the left leg. The tactile sensation was decreased over the lower part of the chest, in which area the pain was also diminished. The muscle and joint sensation in the toes and ankles was affected, and there was a slight nystagmus. The symptoms in this case were considered due to a spinal cord tumor or multiple sclerosis. The spinal fluid after withdrawal showed the changes of Froin's syndrome with many lymphocytes. An operation was performed, and at the sixth dorsal segment two prominences the size of a pea were noted on the spinal cord. When these small elevations were incised, a few drops of clear fluid escaped, and the blebs collapsed on the posterior side of the cord. There was inflammation of the pia mater with little tubercles the size of a pinhead scattered over its surface. On examination of these tubercles, it was found that they consisted of a fibrin network containing a few spindle and stellate cells suggesting, in the mind of the pathologist who examined them, a syphilitic infection.

Case 5 (Launys, Froin and Ledoux[18]).—The diagnosis in this case was meningomyelitis syphilitica. The patient was a laundress, aged 48, who entered the hospital because of difficulty in walking. Four months previous to her admission, she had noted sharp pains in her right ankle shortly after arising in the morning. Within a few days the pain involved the calf muscles and knee of the same leg. The limb became numb, very weak, and walking became almost impossible. The left thigh became involved, and she was able to walk only with support. For one month previous to her coming to the hospital, the lumbar spine had been painful, and her right leg was powerless. During the week previous to her admission to the hospital, the pain had been exceedingly severe.

Examination revealed complete paralysis of the right leg, with the exception of the adductor muscles. Passive movements were painless. The knee jerks were absent on both sides, and neither Babinski's sign nor ankle clonus was

17. Klieneberger, quoted by Mix: Surg. Clin. of J. B. Murphy **4**:317-382, 1915.

18. Launys, Froin and Ledoux, quoted by Mix: Surg. Clin. of J. B. Murphy **4**:317-382, 1915.

present in either leg. Sensation to pin point was very much diminished on the right side, but heat sensibility was normal. There was retention of feces and incontinence of urine. The biceps and triceps reflexes and pupillary reactions all were normal. The condition progressed gradually, the left leg becoming paralyzed; and the patient developed a flaccid paraplegia. The incontinence of urine persisted, and hyperesthesia occurred on the right side. The spinal fluid was withdrawn on four occasions; two fluids coagulated spontaneously and two did not. The spinal fluid Wassermann reaction was negative on all four occasions.

Necropsy in this case revealed a normal spinal cord, except in the lumbar region and in the region of the conus. In the region of the conus terminalis, there was an ovoid swelling, 5 by 6 c.c. long by 3 by 4 c.c. broad, which was soft and fluctuating. The overlying dura mater was intimately fastened to the right surface of the swelling, and slightly adherent to its left surface. The involvement of the subarachnoid space was caused by a fibrous concrescence of the dura and cord on the right side, and by the inflammatory adhesions of the meningeal surface of the left side. The outer surface of the meninges was smooth and apparently normal. There was a small mass which glued a few of the cauda-equina roots together at one point. The examination of the tissue which agglutinated the roots and dura showed it to be meningeal. There was a diffuse radiculomeningitis, with abundant lymphocytes. The arterioles showed a chronic endarteritis and also periarteritis. The general microscopic diagnosis was gummatous meningomyelitis.

Case 6 (Tinel and Gastinel [19]).—A man, aged 35, complained of severe pains in his legs resembling a bilateral sciatica with a later development of weakness. Examination revealed loss of reflexes and changes in his sensibilities. There was paralysis of the legs with a definite Babinski sign at first. During a later examination, plantar irritation caused no response. At first, there was retention of urine, which was later followed by urinary and fecal incontinence.

Necropsy revealed an endarteritis and periarteritis with lymphocytic and plasma-cell infiltration in the cord and meninges. The microscopic findings in this case lead to a diagnosis of subacute meningomyelitis syphilitica.

Case 7 (Souques and Lantuéjoul [20]).—This patient complained of exceedingly severe shooting pains in the legs. There were definite motor and sensory disturbances. The spinal fluid showed the changes typical of Froin's syndrome, with the execption of xanthochromia. The case was considered by the authors as one of meningomyelitis syphilitica.

As previously stated, the etiology of loculated meningitis associated with Froin's syndrome in the spinal fluid is open to question in most cases which have been reported. We believe the following case to be the first in which the diagnosis of syphilis has been definitely confirmed by the demonstration of *Spirochaeta pallida* in the spinal tissues.

Case 8 (Author's Case).—Mr. D. G., colored, aged 25, entered the University Hospital complaining of aching pains in the lower part of the back, slight difficulty in passing his urine and generalized weakness. The family history was negative. The patient had been married for two years. His wife had had one miscarriage, and there were no living children. In the past he had had measles,

19. Tinel, J., and Gastinel, P.: Progrès méd., Paris **28**:377-382, 1912.
20. Souques, A., and Lantuéjoul, P.: Rev. neurol. **36**:137-140, 1920.

mumps, whooping cough, and frequent attacks of tonsillitis and pleurisy. He had a chronic cough and at times night sweats. There had been frequent attacks of nausea, but no vomiting. There was a history of several attacks of gonorrhea and a hard chancre seven years previously, with no antisyphilitic treatment. The present illness had started two months previously with a severe "cold," pains in the chest, and cough, which condition had confined him to bed for two weeks. Following this, he failed to recover his strength, although he had no special symptoms. Two weeks previous to admission, he noticed a persistent aching pain in the lower lumbar region. Within a brief time sharp shooting pains appeared in his legs, and the leg muscles became very sore. Walking became difficult because of the pain, stiffness and soreness in his legs. The weakness progressively increased, night sweats appeared, and he rapidly lost 40 pounds (18 kg.) in weight. The chronic nonproductive cough and pleuritic pains increased. A few days before admission, he had frequency and urgency, which suddenly changed to difficulty, in passing the urine. There was a general headache, especially during the night. There had been some diplopia and frequent epistaxis with dizziness on arising. The feet felt numb and tingled, and he staggered, especially while walking in the dark. His memory remained good.

Examination.—The patient was well developed and nourished. There was considerable discomfort during the examination owing to severe pain on bodily movement. The mentality was clear. The face was pale, and profuse perspiration stood out on the forehead. The pupils reacted sluggishly to light but normally in accommodation. There were no extra-ocular palsies or exophthalmos. The tongue was coated, protruded straight in the midline, and was not tremulous. The cervical glands, especially those in the upper chains, were enlarged. The glands were discrete, soft, hot and tender. Similar glands were present in both supraclavicular fossae. Examination of the chest was difficult, but the lungs and heart appeared to be normal. The abdomen was slightly distended and exceedingly tender, but no abnormal masses were felt. The abdominal respiratory movements were limited. The splenic edge was felt just below the costal border. All the superficial glands of the body were enlarged and of similar nature to those in the neck. Pressure over all the bones caused severe pain, which was also induced on manipulation of the joints. The spine was held rigid, and the spinal muscles were tense and tender. The neck was slightly rigid, while the head was slightly retracted.

Feb. 14, 1923: The neurologic examination on admission showed the biceps and triceps reflexes to be normal, with no paralysis of the arms. There were no atrophies or deformities. The abdominal reflex was present. The knee jerks were present but much diminished. The Achilles reflexes were present and plantar irritation caused flexion.

Course of Illness.—Feb. 17, 1923: The patient felt well if he were not touched. The headache was relieved. He was unable to move either leg. There was retention of urine and severe pain on passive movement of the legs, the patient being apprehensive. The arms were used normally, although there was an occasional twitch. The face and tongue showed no palsy. The head was moved freely and painlessly except in flexion. Examination of the fundi revealed choked disks on both sides, and the veins were dilated. There were no muscular contractions. Both the knee and Achilles jerks were absent, while the biceps and triceps were normal. The plantar response was absent, although the patient felt the "tickle." Deep and superficial pain sense was present in

both legs, while the sensation of motion and position was impaired in the toes of both feet. The sensation to light touch was impaired in both feet but present above the ankles; the outline, however, could not be obtained.

Feb. 18, 1923: The patient was more stupid and sleepy, and found it difficult to hold his eyelids open. The optic disks were slightly less swollen. There was no aphasia present. The great toe on each foot could be moved slightly. The plantar reflex was absent, as were also the knee and Achilles jerks. The sensory disturbances were the same as on previous examination. The urinary retention was still present.

Feb. 19, 1923: The patient was lying on his side, owing to pain in the back, and was more stuporous. On being aroused, he was confused. The head was retracted, and the neck was stiff. The pupils were equal, and extra-ocular movements were normal. There was no facial palsy or difficulty in the use of the arms or hands. The sensations seemed normal in the hands. There was an analgesia to pin point up to the waist line. The patient complained of pain when the legs were moved. The urinary retention was still present.

Laboratory Examination.—The urine showed two plus albumin, with 150 white blood cells per low power field and a few finely granular casts, but no red blood cells. The blood showed a hemoglobin of 80 per cent., with 4,480,000 red blood cells and 5,600 white blood cells per cubic millimeter. A differential blood count revealed nothing unusual. The blood pressure was 138 systolic and 84 diastolic.

Shortly after admission, a spinal puncture was performed. The fluid was under normal pressure and flowed freely. The spinal fluid was xanthochromatic, and a few minutes after withdrawal a large pale, transparent veil-like, spherical pellicle floated in the upper part of the liquid. The fluid showed four plus globulin and boiled solid. The cell count was 215 small mononuclears per cubic millimeter. Three days after the first lumbar puncture a second was performed. On inserting the needle between the third and fourth lumbar vertebrae where the first puncture had been performed, no fluid was obtained. Insertion of the needle between the second and third lumbar vertebrae secured a fluid similar in all respects to the fluid first obtained. At the time of the second puncture, a cisterna puncture was attempted, but only blood was obtained. The Wassermann test on the spinal fluid was negative. The colloidal gold curve was 000000122000, while the mastic was 012340. The blood Wassermann test was four plus. One blood culture was negative. Several guinea-pigs were inoculated with the spinal fluid, and after two months were found to be negative for tuberculosis.

One of the enlarged glands removed from the neck for pathologic examination showed gummatous lymphadenitis with areas of caseation and complete loss of lymph gland structure, which had been replaced by a vascular granuloma, rich in plasma cells and mononuclears, with many vascularized circumscribed areas undergoing caseation. There were no giant cells. The pathologist gave a diagnosis of a very active and virulent syphilitic lymphadenitis.

The roentgenogram of the lower spine showed an occult spina bifida of the upper segment of the sacrum. The roentgenogram of the lungs was completely negative.

Interval History.—Shortly after admission, the patient was placed on a treatment consisting of mercury rubs with 250 grains (16 gm.) of potassium iodid daily. This treatment was discontinued after five days. During this period, the glands in the neck became slightly smaller, and the patient's condition was a

little improved. His temperature averaged 101 F., with two brief remissions to 99 F. During the first five days, his pulse rate averaged 100, when it suddenly rose and remained at an average rate of 130. The pulse fell, and the temperature rose shortly before death.

Necropsy.—The body was that of a young adult negro, showing poor nutrition. The pupils were circular but unequal. On the right side of the neck were several large nodular masses, chiefly in the upper part of the neck and along the posterior border of the sternocleidomastoid muscle. These were evidently enlarged lymph glands, the upper ones being soft and fluctuant, while the lower ones were firmer and smaller. There was a slight protuberance in the lower part of the abdomen. There was a small area of sacral decubitis. The mucous membranes were cyanosed, and the panniculus was scanty.

The spinal cord was examined by the anterior incision, and removed unusually high in the cervical region for this method. The vertebrae cut with unusual difficulty. The dura was found to be slightly more adherent than normal to the inner meninges. In Figure 1, which is a drawing made shortly after the removal of the cord, the incised area is shown rolled back and its inner surface covered with a diffuse granular layer, which was apparently new-formed tissue. The lower meninges of the cord were everywhere diffusely thickened, opaque, and the normal markings were entirely smoothed out. As revealed by the drawing (Fig. 1), there was no definite dural adhesion or loculation of the cord into an upper and lower segment. While the surface of the meninges appeared granular, no discrete tubercles were seen. There appeared at intervals in the upper meninges of the cervical and thoracic cord, hemorrhagic areas which were of considerable size. These were rather diffuse, and in the state examined did not appear to produce elevations or blebs on the surface.

The skull was very thick. The dura tense and more opaque than normal. The meningeal vessels all showed severe congestion. The basal vessels were also congested and enveloped by a diffuse increase of stroma and fibrosis. The inner meninges were congested, opaque, and showed small opaque patches up to 1 mm. in diameter, the larger of which showed slight caseation. The smaller plaques were gray, rather flat, and not sharply demarcated. At the base, the meninges were much thickened, and the opacity was of greater degree. The sectioned brain showed only slight congestion and edema. The ventricles and ependyma were normal. The choroid plexus in the region of the pineal body showed a diffuse fibrosis.

There was a slight collection of bloody fluid in the left pleural cavity. There were numerous enlarged lymph glands in the mediastinum. The thymus was present, hyperplastic and measured 3 cm. in breadth and 0.5 cm. in thickness. The heart and coronary vessels showed no changes. There was a purulent bronchitis in the left lung. The right lung showed severe congestion and edema at the base, with a more pronounced purulent bronchitis which resembled a diffuse and fibrinopurulent lobular pneumonia. The bronchial nodes were enlarged and showed areas of caseation. The thoracic portion of the aorta showed throughout its entire length a marked aortitis in the form of slightly elevated, yellowish ridges, which ran transversely.

The spleen was small, and on section no tubercles were found. There was a universal adhesive perihepatitis. The subdiaphragmatic peritoneum showed numerous irregular thickened plaques, some of which resembled tubercles. On section of the liver, no tubercles or gummas were found, The mesenteric

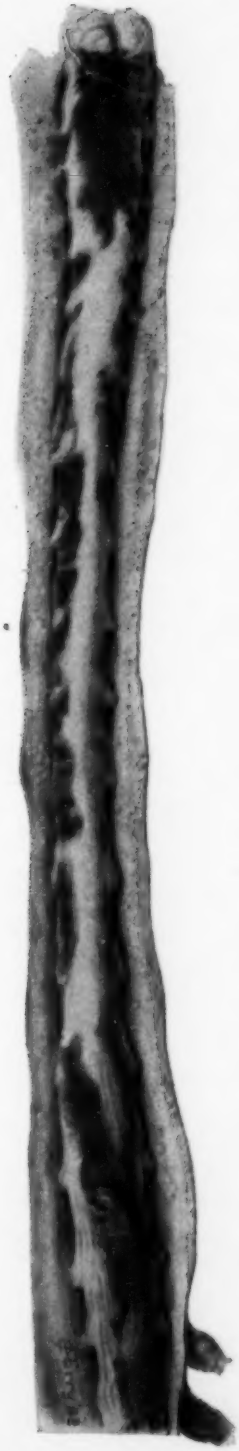

Fig. 1.—The spinal cord of patient in author's case.

lymph nodes were enlarged, and showed areas of caseation. The abdominal aorta was negative. Both kidneys showed moderate parenchymatous changes.

The left testes showed almost complete fibrosis. The bladder was much distended.

Microscopic Findings.—The brain showed a diffuse syphilitic meningo-encephalitis, with edema, congestion and many areas of perivascular plasma cell infiltration.

The meninges showed an older chronic syphilitic meningitis with fibroblastic areas and fibroid thickenings. The section shown in Figure 2 is from the inner border of the spinal dura, and shows syphilitic granulation tissue. Throughout the older process were many areas of active gummatous arteritis.

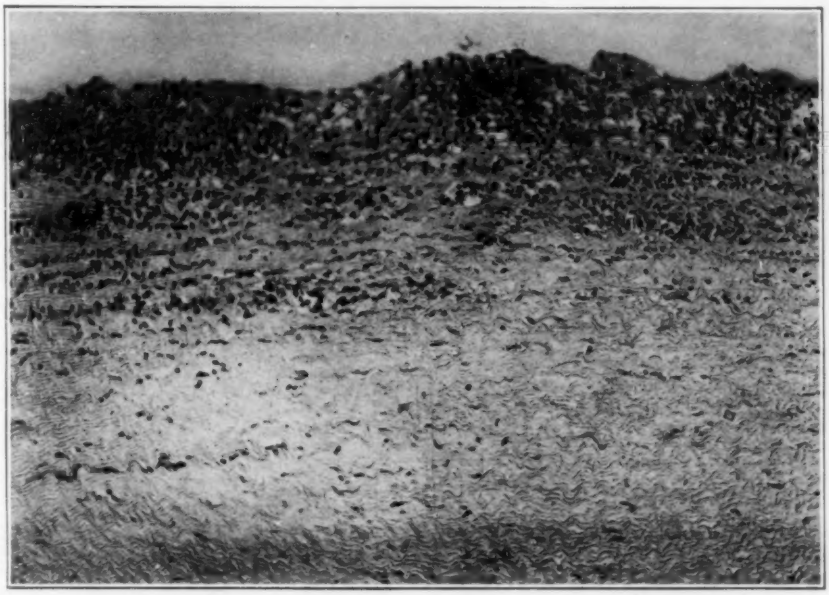

Fig. 2.—Section from the inner border of the spinal dura showing syphilitic granulation tissue.

The vessels showed plasma cell mantling, proliferation of capillaries, proliferating endarteritis, endophlebitis with organizing thrombi, and, as revealed in Figure 5, typical perivascular syphilitic infiltration. There were distinct gumma formations, as demonstrated in Figure 3, which shows a small early gumma on the inner surface of the spinal dura. It also was in this same section that *Spirochaeta pallida* was demonstrated. The cord presented a meningomyelitis. As previously mentioned, and as shown in Figure 1, there were in the cervical and thoracic regions of the cord hemorrhagic areas in the inner meninges. In the state examined, these areas did not appear to form blebs, but in Figure 4 is shown a fluid containing space in this hemorrhagic fibrinous exudate of the inner meninges. The aorta showed syphilitic mesaortitis with one portion showing a gummatous aortitis, while small gummas, as shown in Figure 6, were present and developing in the spinal nerves, which probably accounts for the severe radicular pain that the patient suffered.

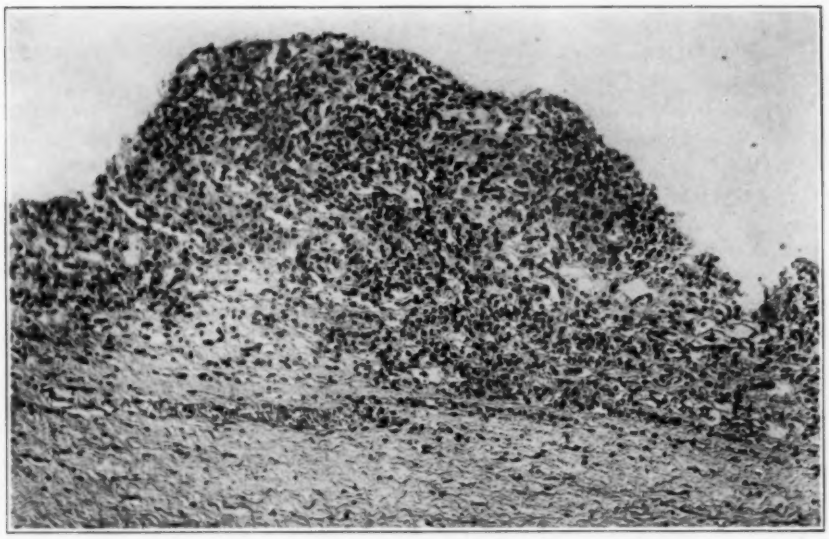

Fig. 3.—Section showing a small early gumma on the inner surface of the spinal dura.

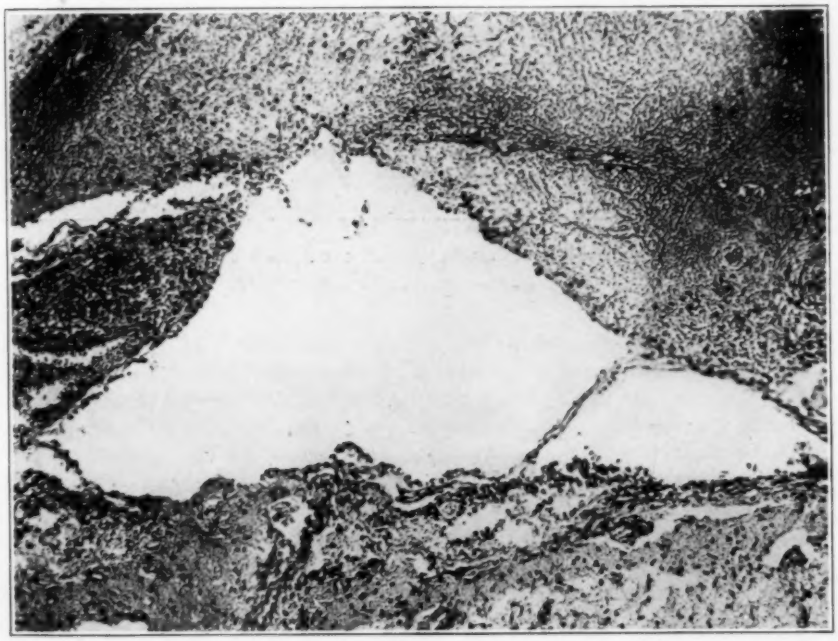

Fig. 4.—Section showing spaces containing fluid in the hemorrhagic fibrinous exudate of the inner meninges.

The lungs showed one small area of tuberculosis, with a small cavity, also a bilateral hemorrhagic purulent bronchopneumonia. The bronchial nodes showed old calcareous, caseating and fibrous tubercles.

The liver was of the extreme nutmeg type. Some of the island of Glisson showed a lymphocytic infiltration, while others showed a cirrhosis of the syphilitic type.

The peripheral lymph nodes all showed gummatous lymphadenitis.

There was active syphilis of the testes.

In the section of the spinal cord meninges shown in Figure 3 *Spirochaeta pallida* was demonstrated by the silver-agar method. In Figure 7 is shown a group of spirochetes occurring in the granulation tissue on the inner surface

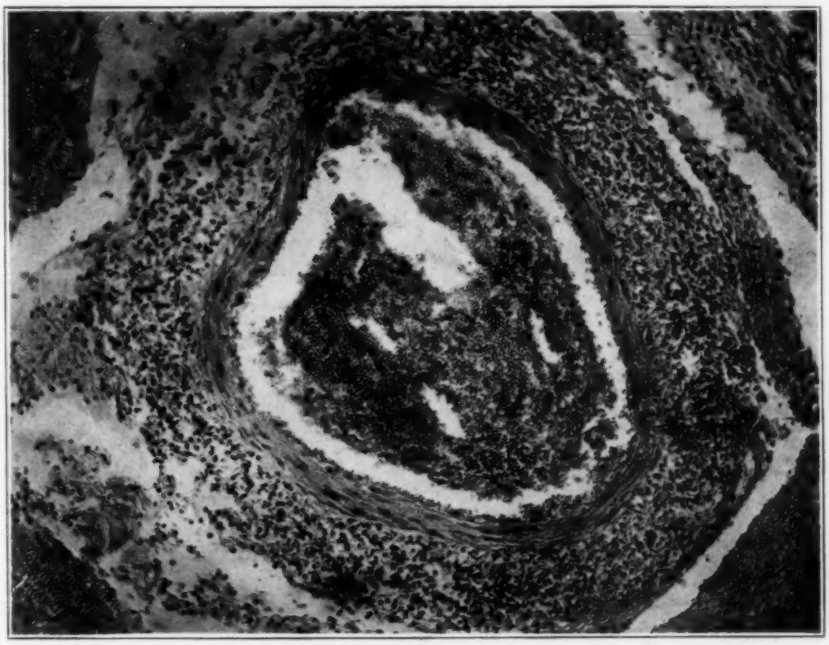

Fig. 5.—The meningeal vessels showing perivascular syphilitic infiltration.

of the dura. It evidently is impossible to get a single spirochete into one focal plane, and several pictures were taken at different focal depths, four of which are here presented in order to show the characteristics of the organism. The spirochetes were easily recognized beneath the microscope, but are difficult to reproduce.

COMMENT

Since attention was first drawn to "cloisonné" or loculated meningitis associated with the syndrome of Froin in the spinal fluid, the question of the former's etiology and the pathologic changes necessary for the production of the latter have been widely discussed.

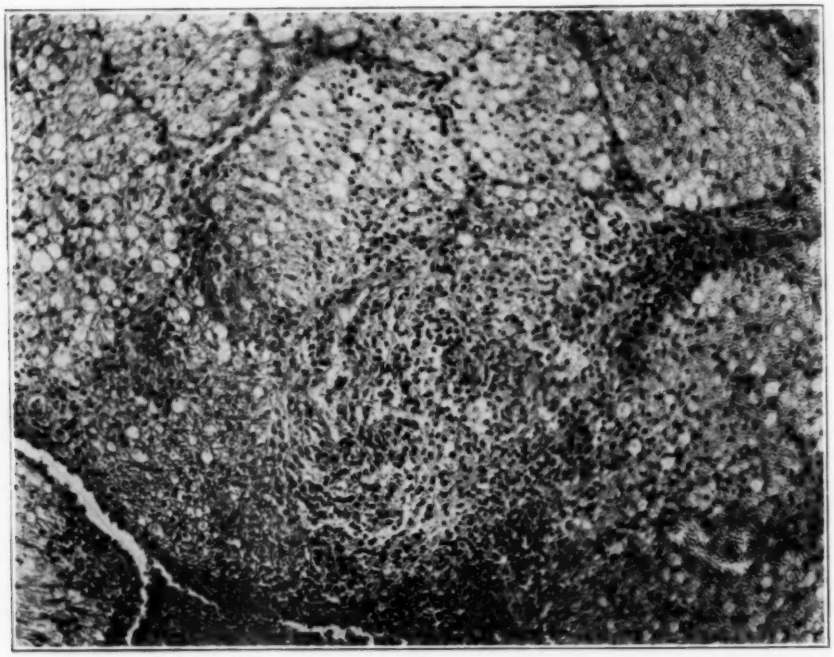

Fig. 6.—Small gumma developing in a spinal nerve.

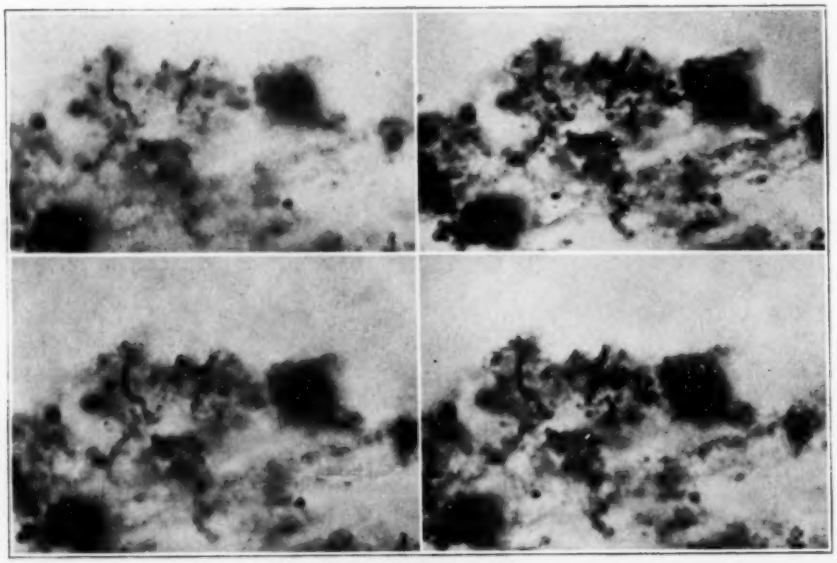

Fig. 7.—A group of Spirochaeta pallida in the syphilitic granulation tissue of the meninges of the inner surface of the dura. As it is impossible to get a single spirochete into one focal plane in granulation tissue, such as here demonstrated, the several pictures have been taken at different focal depths in order to show the characteristics of the organism.

While working with cats, Ayer [21] produced compression of the spinal cord by the insertion of melted paraffin. Necropsy on these animals showed the paraffin to be closely molded over the dorsal aspect of the dura to a depth of 1 to 3 mm. and extending along the dura for a distance of 5 to 9 cm. The cerebrospinal fluid above the compression was normal in all respects, while below was found fluid showing the changes typical of Froin's syndrome. At times vascular engorgement of the pial vessels occurred below the obstruction. Ayer [21] believed that the pathologic process operative in the formation of this changed fluid, and especially the formation of fluids rich in protein, was transudation into the lower sac.

The changes were considered by Hanes [22] as being due to localized obliteration of the pia-arachnoid space, with the formation of a cul-de-sac. He found that when meningitis was not present there was no pleocytosis, and considered the presence of pleocytosis as strongly suggestive of either syphilis or tuberculosis. Hanes likewise believed that the Froin-Nonne syndrome when present always indicated a compressive lesion of the cord.

In Mestrezat's [23] monograph on this subject, he presents two factors which he considered absolutely essential. These are: (1) the formation of a lumbar cul-de-sac which is cut off from communication with the fluid in the upper part of the cord either by meningitis, tumor or disease of the bones of the spine, and (2) the engorgement of the spinal veins below the level of the block, or alteration of the vessel walls by the inflammatory process.

As stated by Greenfield, the degree of change in the fluid depends more on the completeness than on the nature of the block, but certain constituents of the fluid may vary in relation to the nature of the obstruction.

Most necropsies have shown a lumbar cul-de-sac (Raven). However, there have been numerous cases, and especially those resembling Landry's paralysis, or where there has been a rapid clearing up of the spinal fluid changes, in which the idea of an obstruction does not seem tenable. While the theory of a closed cavity appears justifiable in view of the findings at many necropsies, this does not explain cases of polyneuritis or those resembling Landry's paralysis. The rapidity with which patients recovering have shown normal fluids points against inflammatory adhesions in the meninges. A slight edema or swelling of

21. Ayer, J. B.: Cerebrospinal Fluid in Experimental Compression of the Spinal Cord, Arch. Neurol. & Psychiat. **2**:158-164 (Aug.) 1919.

22. Hanes, I. M.: Am. J. M. Sc. **152**:66-71, 1916.

23. Mestrezat, W.: Le liquide céphalo-racidien normal et pathologique, Paris, 1912.

the cord may explain the changes in these cases. The case presented did not show the formation of a cul-de-sac. The meninges were slightly more adherent than normally but, as is clearly shown in the drawing (Fig. 1), there was no annular constriction or adhesion; but the spinal fluid withdrawn from the lumbar region showed the syndrome of xanthochromia and massive coagulation with a lymphocytosis.

The etiology of cases of meningitis showing the syndrome of xanthochromia and massive coagulation in the spinal fluid usually has been doubtful. The meningococcus and staphylococcus have been isolated, but the great majority has gone unproved. In most cases, syphilis or tuberculosis is considered but rarely verified, as associated syphilis does not necessarily prove the etiology. The ultimate test rests on the effect of treatment or on the pathologic demonstration in the tissues. The case of Babinski is the only one found which showed improvement under antisyphilitic treatment. Many persons have shown tissue changes at necropsy which strongly suggested syphilis. In our case, the microscopic evidence of syphilis involving the cord, meninges and spinal nerves is demonstrated in the several photomicrographs; but we also have for the first time conclusively demonstrated *Spirochaeta pallida* in the spinal tissues in a case of so-called loculated meningitis.

SUMMARY

A brief review has been given of the syndrome of Froin with the various views concerning its formation and etiology.

Several cases have been presented which were of probable or proved syphilitic origin.

A case of loculated meningitis in the tissues of which were demonstrated *Spirochaeta pallida* has been presented.

Attention has been drawn to the fact that this case did not show the formation of a cul-de-sac in the spinal canal to explain the formation of the xanthochromia and massive coagulation in the spinal fluid.

PATHOLOGY AND PATHOGENESIS OF UNILATERAL INTERNAL HYDROCEPHALUS *

REPORT OF A CASE

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Unilateral hydrocephalus is a unique condition, the more so when limited to a portion of one ventricle. By unilateral hydrocephalus is ordinarily meant a condition that results from obstruction at the foramen of Monro. It differs but little from the bilateral type, except that in the latter the obstruction is lower down. The mechanism of these obstructive forms has been shown by Dandy.[1] But when the condition is unilateral and no obstruction is present, further study must be made to determine its cause. Such a study forms the basis of this report.

REPORT OF CASE

History.—P. D., a man, aged 62, was admitted to the neurological wards of the Philadelphia General Hospital, Oct. 16, 1921, complaining of paralysis of the right arm and leg, inability to control the bladder and bowels, and difficulty in talking. No history could be obtained from him because of his lack of knowledge of English, plus aphasia. The history given by his daughter was that he had always been intensely alcoholic. Four years prior to admission to this hospital he was found paralyzed on the right side and unable to talk.

Examination.—On admission, he had a right hemiplegia with motor aphasia, and definite evidences of a senile deterioration. With the exception of a little flatness of the lower part of the right side of his face, his cranial nerves were normal, although his visual fields could not be tested. There was a marked spastic paralysis of the right side, with flexor contractures, more marked in the upper extremity. Ankle clonus and the Babinski sign were present on the right. Sensation could not be tested. He was incontinent. His blood pressure was: systolic, 180; diastolic, 110. The diagnosis made at this time was an old thrombotic lesion in the region of the internal capsule.

Course of Illness.—He gradually became stronger and was sent to a convalescent home, from which he was readmitted to the hospital on March 9, 1923, in semistupor, with even greater spasticity than before. His Babinski sign

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* From the Neuropathological Laboratory and Neurological Wards of the Philadelphia General Hospital.

1. Dandy, W. E.: Experimental Hydrocephalus, Ann. Surg. **70:129:** (Aug.) 1919.

was more evident. It was believed that he had had another "stroke." Three days after admission he suddenly became much worse and died, supposedly from another "stroke." His laboratory findings, including the spinal fluid, were negative, except for the evidences of a chronic nephritis.

Necropsy.—The important gross findings were: generalized arteriosclerosis, chronic interstitial myocarditis and chronic tubular nephritis.

The brain on removal was soft, with fluctuation in the left parieto-occipital region and recent areas of thrombotic softening in this area, flattening of the convolutions and a generalized shrinkage of the left side. The vessels were sclerotic; the pia-arachnoid was but slightly thickened. On hardening, the

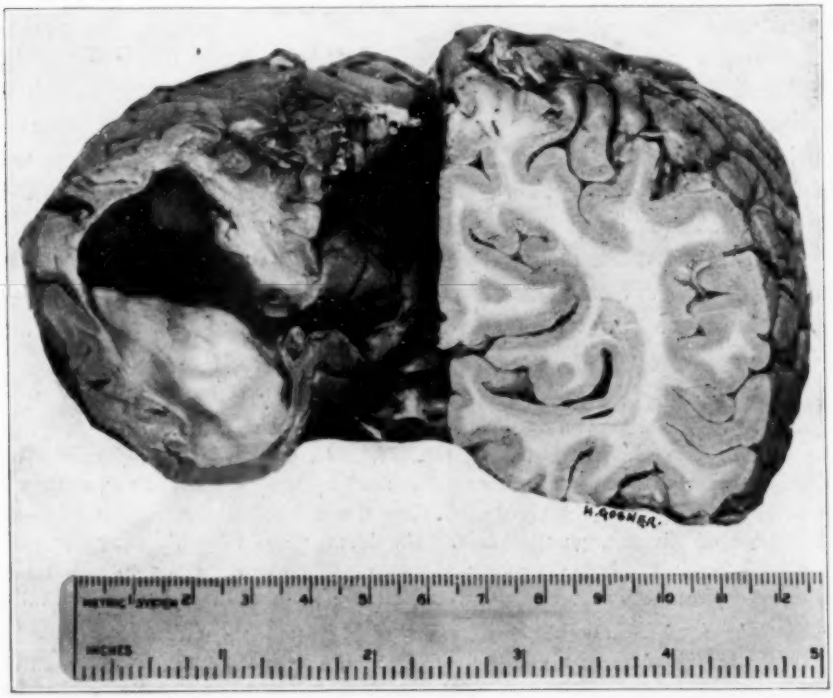

Fig. 1.—Section through the occipital lobes, showing normal right posterior horn and marked dilatation on the left, with thinning of the walls and general shrinkage of the left hemisphere.

brain was cut in frontal sections for a complete study. The right ventricular system was dilated as much as is usually found in a senile brain (Fig. 2), with marked dilatation of the left posterior horn and thinning out of its overlying cortex, as shown in Figure 1. No actual obstruction was found, although the foramen of Monro was slightly narrower than the opposite side. The anterior portion of the left ventricular system and the descending horn were more dilated than the corresponding parts on the right (Fig. 2), but not nearly so much as the posterior portion.

Microscopic Examination.—Sections were studied from all parts of the cortex on both sides. The characteristic findings of an arteriosclerotic brain were present on the right and also in the anterior part of the left hemisphere. It is in

the posterior portion of the left hemisphere that the chief interest lies. Here the cortex was much narrowed and was so altered as the result of softening as to be unrecognizable. In places there was complete replacement by a glial scar. In some portions the replacement was not so complete, and in other areas the process was fairly recent, with the destructive process still going on, as was evidenced by phagocytic cells within the tissue and in the perivascular spaces. The ependymal cells lining the left posterior horn were altered and few in number (Fig. 3). The pia-arachnoid varied in its structure, correspond-

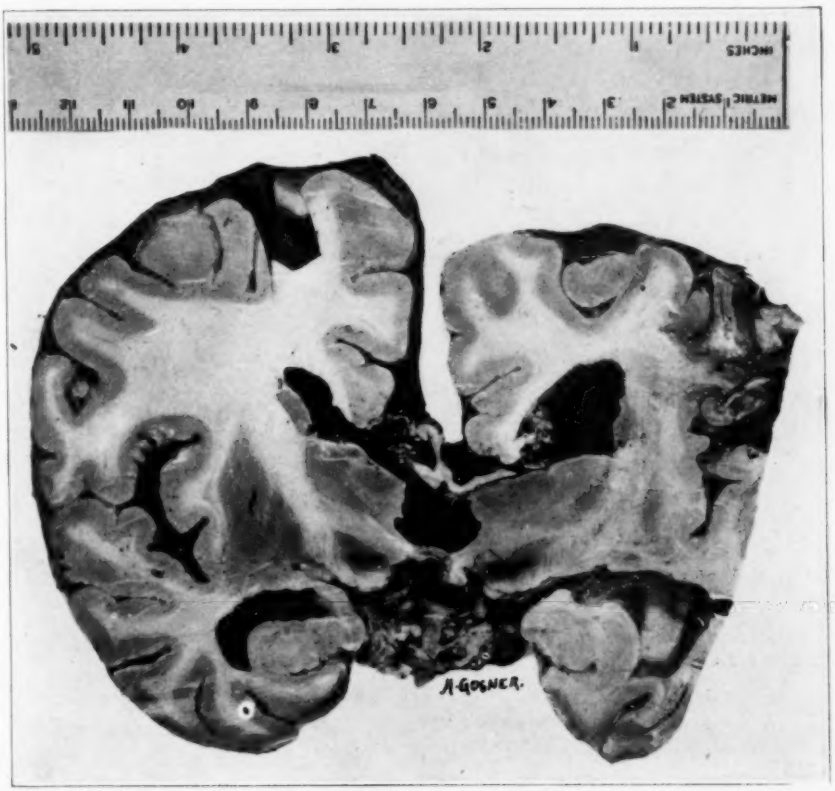

Fig. 2.—Slight dilatation of right ventricle (left side of picture) and great dilatation of left ventricle, with shrinkage of left hemisphere. (N. B. extreme right of picture spoiled in developing.)

ing to the pathology beneath. Over a comparatively "normal" portion, it showed a moderate fibrosis without cell infiltration, while over the posterior horn of the left ventricle, it showed both inflammatory and proliferative changes. In places, there was an adherence of the cortex to the pia-arachnoid, with projection of glial fibers into the pia-arachnoid in the form of sheaves, the cell bodies remaining in the outermost zone of the cortex (Fig. 4), similar to that found by Globus[2] in his case of porencephalus.

2. Globus, J. H.: A Contribution to the Histopathology of Porencephalus, Arch. Neurol. & Psychiat. **6**:652 (Dec.) 1921.

The blood vessels, as is usual in senile cases, varied in structure in different parts of the brain. In places, they were comparatively normal, and in other places, they showed a marked intimal hyaline thickening with resulting narrowing of the vessel lumen. The left pyramidal tract showed both sclerosis and atrophy. The choroid plexus contained masses of pigment and fat, but an equal amount on the two sides.

REVIEW OF LITERATURE

The literature on unilateral hydrocephalus is not abundant. It is first mentioned by Von Mohr,[3] wherein he describes two cases.

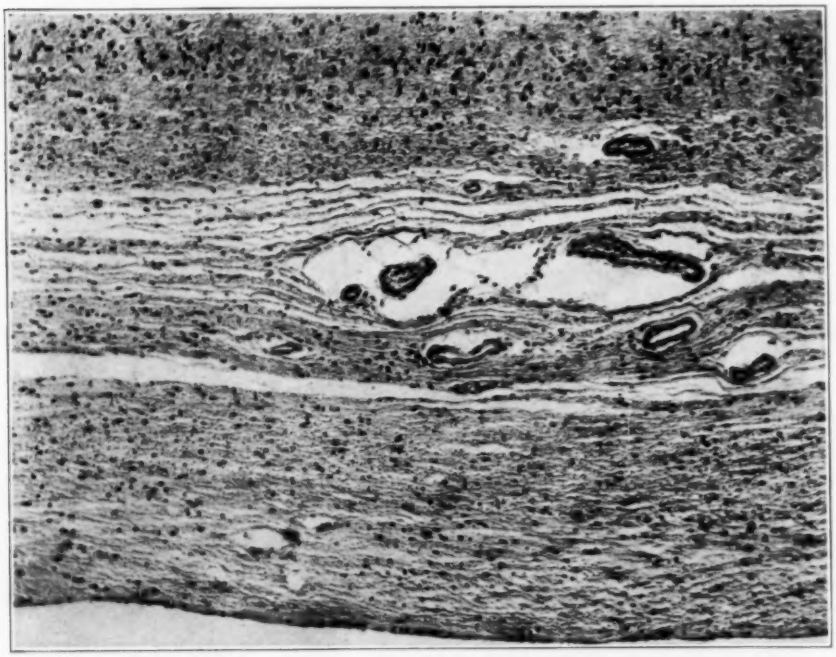

Fig. 3.—Deeper layers of cortex showing altered ependymal cells (at bottom) and evidences of tissue destruction throughout. Hematoxylin-eosin stain; × 69.

Case 1.—A girl, who had been defective and in addition epileptic from her tenth year, had had a right hemiparesis from infancy. She died at the age of 21 of an intercurrent infection. Necropsy revealed a large left ventricle, with a nearly normal right one. There was atrophy of the left hemisphere and also of the left optic nerve. There was some enlargement of the opening to the left descending horn. The ependyma of this ventricle was glistening and thick, and the left corpus striatum showed degenerative processes. The wall of the left hemisphere was very thin. No obstruction at the foramen of Monro could be made out.

3. Casper's Wchnschr., 1842, p. 121.

CASE 2.—A woman, dying at the age of 61, from erysipelas, had been an inmate of an asylum for mental diseases for some years. She showed a right hemiparesis, which had existed for many years. There was contracture of the right arm and leg. Necropsy revealed the left lateral ventricle much enlarged and the left hemisphere wall thin and compressed. The right ventricle was normal, with no obstruction at the foramen of Monro. There was marked degeneration, probably cystic, of the posterior portion of the left corpus striatum, with enlargement of the descending horn. The left anterior quadrigeminal body was atrophied.

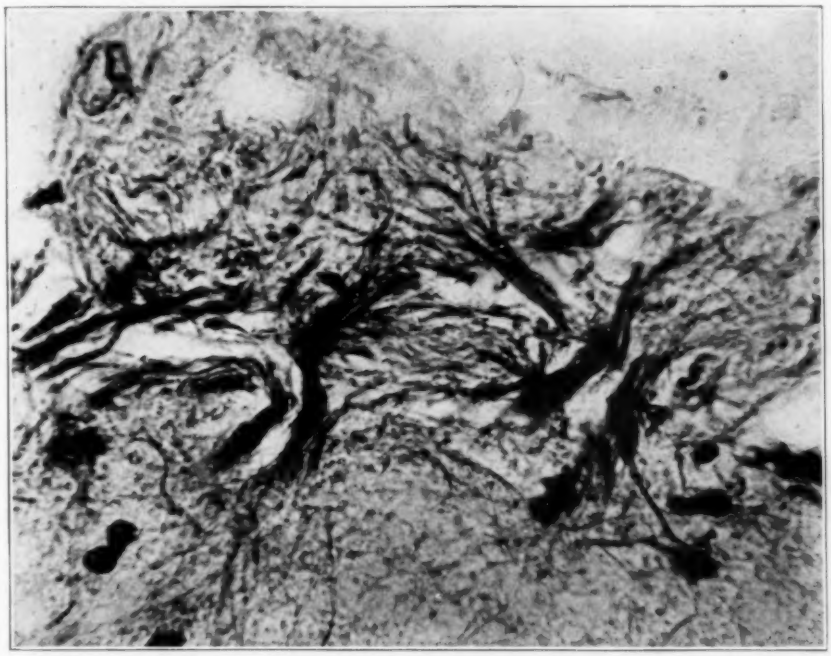

Fig. 4.—Glia cells sending their processes into the pia-arachnoid, which is adherent to the cortex at this point. Phosphotungstic-acid-hematoxylin stain; × 960.

Anton [4] made a careful study of hydrocephalus, especially in the fetus and in children, and came to the conclusion that lessened resistance of the hemisphere wall is responsible in most cases, particularly if weakened by pathologic processes within the walls, and also stated that invariably the dilated ventricle was on the side in which the disease process was located. He likewise found that obstruction would cause the condition.

4. Anton: Zür Anatomie des Hydrocephalus und des Gehirndrüches, Wiener Yahrbuch, 1888.

White's [5] case was that of a woman, aged 74, with a history of repeated hemiplegic attacks of short duration. The limbs on the paralyzed side were less in circumference than the opposite. Necropsy revealed internal unilateral hydrocephalus on the left side. The left choroid plexus showed a large mass, quite firm, containing numerous black spots, which on cross section proved to be hyaloid bodies. The wall of the left ventricle was thick, the ependyma being made up of twelve to fourteen layers of cells. There was marked poverty of cells

Fig. 5.—Section of cortex showing evidences of fairly recent degeneration. Toluidin blue stain; × 69.

in the cortex on that side, also increase of glia and other degenerative changes. The left foramen of Monro was clear. White felt that the condition was one of obstruction due to the mass in the choroid plexus.

A case of unilateral hydrocephalus reported by Spiller [6] was that of an inmate of a school for the feebleminded, of whose family history little was known. She had shown a peculiar gait, with left-sided weakness. She was an idiot and mute. She died at the age of 14, having

5. White, W. C.: Case of Internal Unilateral Hydrocephalus, with Recurrent Attacks of Hemiplegia, Am. J. Insan. **58**:503, 1901.

6. Spiller, W. G.: Two Cases of Partial Internal Hydrocephalus from Closure of the Inner Ventricular Passages, Am. J. Med. Sc. **124**:44, 1902.

had convulsions before death. At necropsy, her right lateral ventricle was dilated, and the cerebral hemisphere on this side was thin, really a mere sack. The left hemisphere was normal. The choroid plexus at the right foramen of Monro was much atrophied and contained numerous masses of cells. Apparently there were nodules of similar structure in the ependyma in the vicinity of the foramen. The left foramen of Monro was nearly normal. The cause given in this case was partial closure of the right foramen of Monro from inflammatory changes about the foramen.

Ziegler [7] reported that there had been found a few cases of unilateral hydrocephalus with closed foramen of Monro, but unfortunately we are unable to find any reference given.

Fig. 6.—The width of the left occipital cortex as shown by Weigert stain; × 45.

Cramer [8] reports two cases, neither of which showed complete dilatation of one ventricle, but only of parts of the ventricle.

Case 1.—An idiot, who died at the age of 20 of anemia, had been epileptic since his seventh year, and had a large head. He also had a spastic right hemiplegia. The left side was underdeveloped. He had difficulty in eating and swallowing. Necropsy showed marked chronic leptomeningitis, which was especially marked over the left frontal and temporal lobes. The left anterior horn was very large and the brain tissue over it so compressed that the hemisphere wall was very thin. As a result of this left frontal meningo-encephalitis, the author explains the consequent shrinking of the wall of the frontal lobe, permitting the anterior horn to enlarge.

7. Ziegler: Lehrbuch der Speciellen Pathologischen Anatomie.

8. Cramer, A.: Lokal beschränkter Hydrocephalus und seine klinische Folgen, Monatschr. f. Psychiat. **17**:561, 1905.

CASE 2.—A man, aged 25, had been ill for three months, with symptoms of brain tumor, such as vomiting, slow pulse, headache, dizziness, and later convulsions. A decompression was performed on the right side, with no relief. There was right hemiparesis and also weakness of the left half of the body. Postmortem examination revealed the right temporal lobe much enlarged, and on opening it considerable fluid escaped. The descending horn was greatly enlarged and was considered by him as cystic. Examination as to the cause showed an inflammatory mass with adhesions at the entrance to the horn, which had entirely closed off the entrance to the horn. While tubercle bacilli were not found, pathologically the mass had the appearance of tuberculous tissue.

Weber,[9] in an extended study into the cause of hydrocephalus in general, including the unilateral type, cites a number of cases, none of which is purely unilateral. No obstruction at the foramen was found in any one of them. In the first case, there was chronic diffuse leptomeningitis with cystic degeneration of the wall of the hemisphere, resulting in atrophy of the brain tissue over the anterior horn. In his second case, there was subcortical encephalitis with softening of the wall, particularly over the dilated ventricle, which was probably syphilitic in origin. The third case showed encephalitis and softening, especially in the hemisphere about the dilated ventricle. Case four showed chronic diffuse leptomeningitis, with adventitial overgrowth and sclerotic areas, especially over the left lateral ventricle. This was probably a case of congenital syphilis. In the fifth case, there was also chronic leptomeningitis, with perivascular infiltration, plus areas of gliosis and softening on the side of the dilated ventricle.

His belief is that the chronic leptomeningitis blocks somewhat the absorption of the spinal fluid, causing some enlargement of both ventricles, but that the greatly enlarged ventricle, amounting to a unilateral type of hydrocephalus, is the result of disease of the hemisphere wall, either inflammatory or sclerotic in origin, resulting in atrophy and contraction of the wall, which permits the ventricle to expand or enlarge—hydrocephalus ex vacuo.

Hunt [10] cites the case of a woman, aged 78, who, twenty-five years previously had had left-sided weakness, from which she never fully recovered, but she could walk. No statement is made as to the presence or absence of blocking at the foramen of Monro, and no explanation of cause is given.

Dandy [1] states that unilateral hydrocephalus is produced by occluding one foramen of Monro. He performed experiments on dogs by blocking one foramen, which was followed by dilatation of the ventricle on that side. He then removed the choroid plexus on that side and

9. Weber, L. N.: Ueber erworbenen Hydrocephalus, Arch. f. Psychiat. **41**:64, 1906.

10. Hunt, E. L.: Unilateral Hydrocephalus, Med. Rec. **89**, 1916.

obtained a complete occlusion of the ventricle. As a result of these findings, he felt that he had proved that the spinal fluid had its origin from the choroid plexus and that the ependyma was not concerned in the production of the fluid.

He reports a case under his observation in which a pedunculated glioma was suspended in the left ventricle and periodically closed the left foramen of Monro. Postmortem examination showed the affected ventricle greatly enlarged and the foramen of Monro completely filled by the tumor, which had been forced into it. Part of the tumor could be dislocated, leaving the foramen only partially occluded. His opinion of the intermittent attacks was that the foramen was patulous during the cessation period.

An analysis of these cases shows that only in three of them was there partial or complete blocking at the foramen of Monro. In the others there was no obstruction found at the foramen, but there was a degenerative process of the hemisphere on the affected side, consisting either of a chronic leptomeningitis, an old encephalitis, or vascular disease of the hemisphere. In two cases there was merely a local enlargement of one horn and part of the body of the ventricle due to inflammatory processes, followed by contraction of the hemisphere wall, and permitting the ventricle or horn to enlarge. In one case the cause is not given, but from notes of the case it would appear that a marked arteriosclerosis was present with degeneration of the hemisphere wall.

Another point of interest in reviewing these cases is found in the great variation in ages, the youngest patient being 14 and the oldest 78. Furthermore, all but two patients had had convulsions, either general or unilateral, and all showed weakness of the body on the side opposite the dilated ventricle. In only two, did the question of syphilis arise.

COMMENT

From a clinical standpoint, unilateral hydrocephalus has not been of great importance because of the difficulties of diagnosis and the rarity of the condition. With such modern methods as the use of Dandy's ventriculography, the condition might be diagnosed, if suspected. From a review of the literature and the study of this case, it is felt that certain symptoms are suggestive of the condition, although nothing is pathognomonic. It is to be considered: (1) when there are either repeated "strokes" on the same side, with or without convulsions or a sudden onset of hemiparesis with progression to complete spastic paralysis over a period of months or years; (2) if slow mental deterioration has been present, especially in combination with the foregoing symptom; (3) or if convulsions have been a rather prominent symptom.

Pathologically in these cases there is a partial or complete unilateral hydrocephalus, depending on the cause and extent of the lesion. If the

condition is due to an obstruction at the foramen of Monro, there will be a more or less uniform dilatation of the ventricle with practically the same pathology in the cortex as is present in the bilateral obstructive type. If no such obstruction is found, the brain substance, its membranes and vessels must be carefully examined. If there are present degenerative changes in the cortex, as is usually the case, the mechanism in the production of this condition even without increase of pressure in the cerebrospinal fluid on that side can be understood. We feel that if there are extensive areas of softening over a portion of the ventricular cavity, there will be first of all a gradual shrinkage of the brain substance in that area with a compensatory pouching out of the ventricle, producing what Weber so aptly calls hydrocephalus ex vacuo. The development of this condition is probably never sudden, but extends over a long period of time. The causes may be varied and may occur at any period of life. In the embryo, an encephalitic process limited to one hemisphere or even bilaterally may produce a unilateral or a bilateral hydrocephalus in the same manner. In childhood and in middle life, similar encephalitic changes may occur as the result of an infection such as syphilis, tuberculosis, etc. In late adult life arteriosclerosis plays the major rôle, whether brought about by intoxication or infection. We feel that the condition has its counterpart in a vascular aneurysm, where, without rise of local pressure, but with a defect in the vessel wall at a certain part there results a definite dilatation.

The pathology of the obstructive type of hydrocephalus, whether unilateral or bilateral, is quite distinct from that which we are describing.

In the case reported here we feel that the course of events was somewhat as follows: There was, in 1917, an occlusion of a vessel of the left cortex producing an aphasia and hemiparesis. It occurred in the cortex and not in the internal capsule because of the fact that the sections showed no softening in the internal capsule but in the motor cortex itself. Gradually, there was a shutting off of the nutrition to more and more of the cortical tissue and at times rather suddenly when the patient had the symptoms of another "stroke." As a result, there was glial replacement of the cortex with shrinkage and resulting dilatation of the ventricle without increase of intraventricular pressure as shown clinically by the absence of headache, vomiting, etc. The process was especially marked over the parieto-occipital areas, but the motor cortex was also involved, as was shown by the histologic sections and clinical course. That the motor symptoms were old was shown by the degeneration and atrophy of the pyramidal tract.

SUMMARY

1. Clinically, unilateral hydrocephalus cannot be diagnosed with certainty. It is to be suspected in older patients when there are repeated

strokes on the same side or a slowly progressive hemiplegia with convulsions and mental deterioration where other conditions, such as brain tumor, etc., can be ruled out.

2. Unilateral hydrocephalus can occur, as shown by Dandy, from obstruction at the foramen of Monro.

3. It can also occur as the result of arteriosclerotic or encephalitic changes in the brain substance.

4. Partial unilateral hydrocephalus occurs as the result of localized vascular or encephalitic changes resulting in what Weber calls hydrocephalus ex vacuo.

DISCUSSION

DR. WILLIAM G. SPILLER: If hydrocephalus develops early in life, or is congenital and affects the cerebral hemisphere of only one side, as in my case, it is associated with atrophy or hypoplasia of the opposite cerebellar hemisphere. I have had two other cases of partial hydrocephalus. In one of these the hydrocephalus was enormous and was confined to the posterior horn of one cerebral hemisphere.

A COMPARATIVE STUDY OF SYPHILIS IN COLORED AND IN WHITE WOMEN WITH MENTAL DISORDER

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WASHINGTON, D. C.

There is a widespread belief among general practitioners and even among psychiatrists in the South that the vast majority of colored insane patients are syphilitic. With this in mind, a study was made of syphilis in a large group of female colored patients in St. Elizabeth's Hospital (Government Hospital for the Insane) Washington, D. C. The relative proportion of colored to white persons admitted to the service was taken up first. Then data were collected concerning the incidence of syphilis in the two groups. The next two subjects studied were the relation of central nervous system involvement and of definite general paresis to the number of syphilitic persons and to the number of admissions. The data were then reviewed from the point of view of age on admission and length of residence in hospital of the patients that died of paresis. Finally the mental reaction types of a small group of colored and white paretic patients were investigated. The conclusions drawn from these statistics do not, of course, apply to negroes in other parts of the country, as practically every patient studied was a resident of the District of Columbia or the immediate vicinity, and it is commonly conceded that the economic status of the negro in the District is unique (Bevis).

MATERIAL USED

This investigation was based on the case records of 1,000 white and 500 colored women admitted to the female service. These were taken in order of admission counting back from Jan. 10, 1923. It so happened that these 1,500 admissions covered just short of seven years, so that when it seemed desirable to make yearly comparisons, those seven years were used. The cases over the seven year interval and the 1,500 cases do not exactly coincide, so that the figures are not interchangeable. Wassermann tests of the blood serum were made on all persons admitted with rare exceptions when the patient died or was discharged before there was time to take a specimen. In every case in which the Wassermann test was returned positive or in which there were suggestive clinical finidings, a second examination of the blood serum was made and also a spinal fluid examination. Tests were repeated if the serological findings did not seem to accord with the clinical findings. The reported data on the spinal fluid examinations included protein content, cell count, Wassermann reaction and colloidal

gold curve. The ages were taken from the admission papers and were doubtless inaccurate in many cases, particularly in those belonging to the colored group, but the errors are probably not great enough to invalidate the results.

RELATIVE PROPORTIONS OF COLORED AND WHITE PATIENTS ADMITTED

During the seven years, from 1916 to 1922 inclusive, there were 1,614 female admissions. Of these 1,063, or 65.8 per cent., were white, and 551, or 34.2 per cent., were colored. Table 1 shows numbers and percentages for each year. It will be seen from a glance at this table that the percentage of white admissions varies between 59.4 per cent. in 1916 and 70.2 per cent. in 1918. There were, therefore, approximately half as many colored as white women patients admitted in the period under discussion.

TABLE 1.—*White and Colored Female Patients During Seven Years*

Year	Total	White		Colored	
		Number	Percentage	Number	Percentage
1916	227	135	59.4	92	40.6
1917	182	118	64.8	64	35.2
1918	242	170	70.2	72	29.8
1919	233	159	68.2	74	31.8
1920	252	167	66.3	85	33.7
1921	227	151	66.5	76	33.5
1922	251	163	65.0	88	35.0
Average	230.5	151.8	65.8	78.7	34.2

OCCURRENCE OF SYPHILIS

The diagnosis of syphilis was made on clinical findings and a positive Wassermann test of the blood serum, positive findings in the spinal fluid, or both. Of 1,000 white patients, fifty-nine, or 5.9 per cent. were syphilitic, whereas of the 500 colored patients, eighty-two, or 16.4 per cent., were syphilitic. It is interesting to compare this with figures obtained in the male service of this same hospital in 1917 when 50 per cent. of the colored persons admitted during the year were syphilitic (Wender). Hindman, writing in 1915, gives figures so similar to those found in the present study as to be striking: sixteen per cent. colored syphilitic and 5 or 6 per cent. white.

Table 2 gives the data by years. It will be seen that the highest percentage of syphilitic persons, among both colored and white, occurred in 1919, when it reached 27 per cent. in the colored group and 9.4 per cent. in the white. The lowest percentage in the white persons admitted was 3.3, which occurred in 1921, whereas the lowest percentage in the colored was 11.1, which occurred in 1918. A glance at Chart 1 shows that, in spite of yearly variations, the general trend of admissions for

syphilis is downward. This seems odd in view of the fact that every year we are becoming more expert in diagnosing syphilis in its early stages. Possibly it is explained by the fact that natural immunity is becoming more and more active. Also, the public is gradually becoming educated in psychiatric problems and patients suffering with mental illness are being brought to the hospital in the earlier stages of their

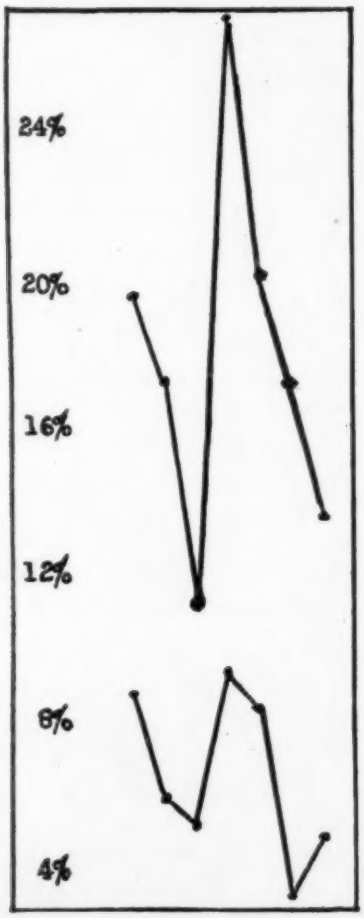

Chart 1.—Percentage of patients with syphilis admitted to the hospital.

disease, thus increasing the total admissions and reducing the proportion of patients with syphilis.

It should be borne in mind that paretic persons should not be classed exactly as other patients in these tables because, whereas others are insane patients who happen to be syphilitic, paretic patients are insane patients *because* they are syphilitic. For this reason paretic patients are discussed as a separate class.

OCCURRENCE OF SYPHILIS OF THE CENTRAL NERVOUS SYSTEM

In collecting data on the subject of syphilis of the central nervous system, only those cases were considered in which the clinical symptoms were confirmed by some positive findings in the spinal fluid. In the present discussion, general paresis is included in this group. Writing in 1917, Bevis says: "The incidence of cerebrospinal syphilis and paresis is relatively low in the Southern negro." Data from this hospital do not support this statement, perhaps because this study is confined to women. Of the eighty-two colored syphilitic women, thirty-nine, or 47.5 per cent., had some form of involvement of the central nervous system. Only twenty-seven of the fifty-nine white patients with syphilis (45.7 per cent.) were so affected.

TABLE 2.—*Showing Percentage of Syphilitic Patients Admitted During Seven Years*

	White			Colored		
Year	Total Admissions	Syphilitic Admissions	Percentage	Total Admissions	Syphilitic Admissions	Percentage
1916.....	135	12	8.8	92	18	19.5
1917.....	118	7	5.9	64	11	17.2
1918.....	170	9	5.3	72	8	11.1
1919.....	159	15	9.4	74	20	27.0
1920.....	167	14	8.4	85	17	20.0
1921.....	151	5	3.3	76	13	17.1
1922.....	163	8	4.9	88	12	13.6

OCCURRENCE OF GENERAL PARESIS

The diagnosis of general paresis was made on the basis of the clinical symptoms, confirmed by spinal fluid findings, regardless of the presence or absence of a positive blood Wassermann test. In many cases a blood Wassermann test which was negative on admission became positive in the course of treatment, and similarly, one which was positive in the beginning became negative. Patients with atypical spinal fluid findings, even though the condition was diagnosed general paresis on the basis of clinical symptoms, are not grouped as such in this study. O'Malley, in 1914, states that although cerebral syphilis is less common in white than in colored persons, the ratio of general paresis is about equal. Conditions have changed to such an extent that this is no longer the case. Thirty-three of the 500 colored patients, or 6.6 per cent., were paretic; eighteen of the thousand white patients, or 1.8 per cent., were paretic. The difference shows up even more clearly when we consider the proportion of paretic to syphilitic patients rather than to total admissions. Thus 40.2 per cent. of colored syphilitic patients are paretic, whereas only 30.5 per cent. of the white patients with syphilis are paretic. Perhaps there is some qualitative difference in the personality of the negro, a stolidity or apathy, which prevents the development of a mental dis-

turbance associated with syphilis until the disease has taken a definite hold on the central nervous system.

Table 3 gives an idea of the yearly variations in paretic admissions, and Table 4 shows the relation of the number of colored patients to the number of white patients. Chart 2 gives the same data in graphic form and would seem to indicate that the proportion of white patients with paresis is on the increase.

AGE OF PARETIC PATIENTS ON ADMISSION

The patients with syphilis were studied in regard to age on admission, and the complete results are shown in Table 5. The only

TABLE 3.—*Showing Percentage of Paretic Patients Admitted During Seven Years*

Year	White			Colored		
	Total Admissions	Paretic Admissions	Percentage	Total Admissions	Paretic Admissions	Percentage
1916	135	2	1.5	92	5	5.4
1917	118	0	0.0	64	3	4.7
1918	170	1	0.6	72	1	1.4
1919	150	6	3.8	74	6	8.0
1920	167	4	2.4	85	5	5.1
1921	151	2	1.3	76	9	1.2
1922	163	4	2.4	88	7	7.1

TABLE 4.—*Showing Relative Proportion of Colored and White Paretic Patients*

Year	Paretic Admissions	White		Colored	
		Number	Percentage	Number	Percentage
1916	7	2	28.6	5	71.4
1917	3	0	0.0	3	100.0
1918	2	1	50.0	1	50.0
1919	12	6	50.0	6	50.0
1920	9	4	44.4	5	55.6
1921	11	2	18.2	9	81.8
1922	11	4	36.3	7	63.7

data of any significance concern the paretic patients. It will be seen at a glance that the colored and white paretic patients correspond as to the age at which the largest number of admissions occur. For instance, the largest number of white paretic patients admitted during any five year age period was five admitted between the ages of 35 and 40. Four were between 40 and 45 on admission. Among the colored paretic patients, seven were admitted between the ages of 35 and 40 and seven between the ages of 40 and 45. The admission age of white paretic patients varied between 25 and 65, whereas the admission age of colored paretic patients varied between 20 and 70. This does not include an 11 year old paretic (colored).

LENGTH OF RESIDENCE IN HOSPITAL OF PARETIC PATIENTS THAT DIED

The average life of colored paretic patients after admission is nine months and twenty-four days. The longest residence was twenty-five months, twenty-seven days, and the shortest was one month, five days. These figures were obtained from the records of seventeen colored paretic patients who died in the St. Elizabeth's Hospital during the past seven years. In the case of nine white paretic patients who died during the same period, the longest residence was thirty-three months and twelve days, the shortest twenty-six days and the average length of life thirteen months and seventeen days. The fact that the families of colored patients are singularly unobservant and do not bring the patients to the hospital until they are in a bad condition may account for this difference.

Chart 2.—Percentage of patients with paresis admitted to the hospital.

TYPES OF MENTAL REACTION FOUND IN PARETIC PATIENTS

A group of paretic patients known to me personally were studied in regard to the general type of mental reaction. There were in this group twenty-four colored women and eighteen white women. In all the cases, deterioration was sooner or later the outstanding characteristic, but an attempt was made to classify them roughly according to the next most conspicuous feature of the mental condition. The differences between the races were marked. Three types of reaction were found in both colored and white patients—euphoria, excitement and confusion. Reference to Table 6 shows that the largest percentage of white patients fell into the group characterized chiefly by confusion. The largest percentage of colored paretic patients, on the other hand, were euphoric. Euphoria was the chief feature of 33.3 per cent. of the white cases and made up the second largest percentage. In 27.7 per cent. of the white patients, excitement was the predominant feature, whereas this was the case in only 8.3 per cent. of the colored patients. Three of the colored

patients were depressed, two in a quiet way, one with periods of apprehension and agitation. It is perhaps worth mentioning that these three colored women had a large percentage of white blood. No white paretic patient showed depression as a prominent symptom. Three of the colored patients showed conspicuous hyperesthesia. They were all confused and unhappy, but pain was the outstanding feature. They had sudden outbursts of crying, apparently associated with pain; they screamed when touched, but were unable to tell in what way they suffered. One of these patients had spasmodic contractions of the

TABLE 5.—*Age on Admission*

Age	General Paresis		Other Cerebrospinal Involvement		Syphilis	
	Colored	White	Colored	White	Colored	White
10-14	1	..	..	..	..	..
15-19	..	..	..	..	4	2
20-24	2	..	..	..	4	3
25-29	2	3	2	..	3	3
30-34	2	2	..	1	8	1
35-39	7	5	..	2	5	6
40-44	7	4	1	2	6	5
45-49	4	1	1	3	3	1
50-54	4	..	1	..	2	2
55-59	..	2	..	..	..	2
60-64	3	1	1	..	1	2
65-69	1	..	..	..	1	1
70-74	..	..	..	1	1	2
75-79	..	..	..	..	2	1
80-84	..	..	..	..	2	..
85-89	..	..	..	..	..	..
90	..	..	..	..	1	..

TABLE 6.—*Conditions in Colored and in White Patients*

	Colored, Percentage	White, Percentage
Euphoria	37.5	33.3
Excitement	8.3	27.7
Confusion	25.0	39.0
Depression	12.5	....
Hyperesthesia	12.5	....
Catatonia	4.2	....

right hand and forearm, lasting two or three minutes at a time and accompanied by excruciating pain. One colored paretic patient showed a reaction differing from any other case studied in that this picture was typical of catatonic stupor. She was mute, negativistic, rigid, tube-fed, and her only reaction to persistent attention was copious lacrimation, the tears coursing over expressionless features.

CONCLUSIONS

1. Over a period of seven years the number of colored admissions to the female service of St. Elizabeth's Hospital was approximately one half the number of white admissions.

2. Of a thousand white patients admitted to the female service, 5.9 per cent. were syphilitic; of 500 colored patients, 16.4 per cent. were syphilitic.

3. During the past seven years the general trend of the percentage of syphilitic admissions has been downward.

4. Some form of involvement of the central nervous system was found in 47.5 per cent. of the syphilitic colored women and in 45.7 per cent. of the syphilitic white women.

5. General paresis was present in 6.6 per cent. of the colored women and in 1.8 per cent. of the white women patients.

6. Of the colored syphilitic women, 40.2 per cent. were paretic, but only 30.5 per cent. of the white syphilitic women were paretic.

7. The most common admission age of both colored and white paretic patients was between 35 and 45.

8. The average length of the life of a patient with paresis after admission to the hospital was found to be less for the colored women than for the white.

9. Confusion was the outstanding feature of about 39 per cent. of the psychoses of the white paretic patients, euphoria in 33.3 per cent. and excitement in 27.7 per cent., whereas euphoria ranked highest in the colored paretic (37.5 per cent.) patients and confusion was the most conspicuous element in 25 per cent.

News and Comment

CHANGES IN EDITORIAL BOARD

Dr. Albert M. Barrett of Ann Arbor, who has been a member of the Editorial Board of the ARCHIVES OF NEUROLOGY AND PSYCHIATRY since 1920, has resigned. Dr. H. Douglas Singer of Chicago has been appointed by the Board of Trustees of the American Medical Association as his successor.

THE AMERICAN PSYCHIATRIC ASSOCIATION

At the annual meeting of the American Psychiatric Association, held in Atlantic City, from June 3 to 6, the following officers were elected for the ensuing year: President, William A. White, M.D., Washington; Vice President, C. C. Floyd Haviland, M.D., Albany; Secretary-Treasurer, Earl D. Bond, M.D., Philadelphia.

Richmond, Virginia, was chosen as the next place of meeting.

Abstracts from Current Literature

MEETING OF THE SOCIETÉ DE NEUROLOGIE DE PARIS. ANDRÉ-THOMAS, *President, July 5, 1923;* Rev. Neurol. **30**:30-86 (July) 1923.

Cerebellar Tumor with Parkinsonian Rigidity and Psychomotor Retardation.—Vincent, Bernard and Darquier presented a woman, aged 77, with a parkinsonian posture of the unusual flexion variety, i. e., with flexion of the lower extremities, who did practically nothing spontaneously (she had to be compelled to eat) and who was blind (double optic atrophy) with eyes deviated to the right. When requested to perform an act she did it two or three times but with increasing sluggishness, and finally stopped altogether. The same was true of answers in conversation. She had no cerebellar signs.

At necropsy a large, easily enucleable tumor the shape of a flattened ball was found in the left cerebellar hemisphere. The cerebellum was merely compressed, not destroyed. The anterior part of the vermis bulged into the fourth ventricle and compressed the posterior quadrigeminate bodies. The aqueduct of Sylvius was much dilated, as were the lateral and third ventricles. The pituitary body was compressed and the foramina of Monro were from two to three times their normal diameter. There was general cerebral edema. The caudate and thalamus appeared to be enlarged. The case was presented to indicate the effect of a cerebellar tumor in producing a parkinsonian picture, the mechanism of which was probably partially dependent on ventricular dilatation and involvement of the central ganglions thereby.

Atypical Striate Syndrome.—Babonneix and Lance presented a child, aged 7½ years, who showed marked developmental retardation and subluxation of both femora. When seen in November, 1922, the child had in addition salivation, strabismus, a speech difficulty, muscular rigidity and athetosis associated with hypotonia at the finger joints. There was also complete reaction of degeneration of both sciatic nerves. Five months later an epileptic attack occurred of twelve hours' duration. In June, 1923, there were present also right central facial paralysis, paralysis of the neck muscles, associated movements in the left arm when the right was voluntarily moved, ventral hernia and mental retardation. There were no changes in the pupillary reactions, no Babinski signs, no sphincter disorder or spasmodic laughing and crying. The blood and spinal fluid could not be examined.

On the basis of benefit from specific therapy the case was regarded as syphilitic. It is not included in any known clinical picture, such as Wilson's disease, pseudosclerosis or Vogt's double athetosis.

Contribution to the Subject of Cervical Ribs and Processes.—Crouzon and Mathieu presented a young woman, aged 23, who complained of severe pain in the left arm. There were double ulnar griffe, atrophy and incomplete paralysis of the intrinsic ulnar muscles of the hand with slowed electrical reactions. There was also mild cervical sympathetic paralysis in the left eye (Claude-Bernard-Horner syndrome).

The hypertrophied processes on the seventh cervical vertebra having been demonstrated by the roentgen ray were removed at operation. They were not ribs. Immediately the griffe (spasm) and pains disappeared. The Claude-Bernard-Horner syndrome did not.

Hypertrophy of both the transverse processes of the seventh cervical vertebra and of the cervical ribs may coexist. The effect is the same. The eighth cervical nerve, as shown at operation, was the one affected.

Inversion of the Achilles Reflex of Spinal Origin.—Souques recalling that he had described this condition in 1911, reported the case of a young man, aged 17, who had a mild spastic paraplegia dependent on an anomaly of the fifth lumbar vertebra and the sacrum. Spina bifida could not be ruled out. Pott's disease was. Souques regarded the deformity as congenital. The patient failed to walk until 3 years of age, and his gait had always been abnormal.

The ankle jerk response (as well as the response from tapping the gastrocnemii) was always dorsiflexion of the foot. Souques believes that a congenital defect in the spinal cord accompanies the vertebral defect.

In the discussion, Sicard observed that he had reported (Rev. Neurol. No. 2, February, 1923) a case of inversion of the Achilles reflex due to a root tumor which cleared up after removal of the tumor.

Probable Striate Syndrome. Spasm of the Face with Tachyphemia, Tachymicrographia and Tachypnoea.—Souques and Blamoutier reported the case of a patient, aged 51, well until four years before, who since had had the symptoms given in the title. When told to write between two lines, micrographia was absent, a phenomenon described by Froment. Epidemic encephalitis was considered, but no definite diagnosis was made failing other confirming evidence.

In the discussion Vincent emphasized the frequency of blepharospasm in idiopathic and postencephalitic parkinsonian syndromes.

Spasm while Reading Aloud with Spasmodic Torticollis, Associated with Synkinetic Movements and Stuttering.—Heuyer and Deyras reported the case of a child, aged 12, an orphan, showing no mental retardation, who was seized with abnormal movements designated as "tics." These slowly disappeared. Six weeks before the patient began to stutter and slowly lost the ability to read aloud. When requested to read the child showed a facial spasm—contraction of the left sternocleidomastoid (torticollis). The right upper extremity showed an athetotic movement—was carried backward, the hand everted and the fingers flexed. A slight tremor coexisted. Rarely, a similar movement occurred on the left. When reading in a low voice these spasms did not occur, nor when reading to himself. Writing was normal. When a question was brusquely asked this train of movement occurred to a slight degree, but disappeared quickly. The patient was undersized, had curved femurs, hypospadias, dental malformations and an arched palate. The Wassermann test was negative.

The etiology was considered as constitutional or congenital emotional instability.

Lumbar Syndrome with Xanthochromic Reaction in the Spinal Fluid Due to a Root Neuroglioma. Diagnosis by Lipiodal Test and Roentgen Ray.—Sicard and Laplane emphasized the need of operation in these cases. Two of the three patients had been operated on, with cure of the symptoms and disappearance of the xanthochromic reaction. The value of the lipiodal test is emphasized.

A Special Type of Pyramidostriate Syndrome Occurring in the Adult. Progressive Spastic Paraplegia. Peribuccal Intention Tremor.—Foix and Valière Vialeix, reported a case in which a patient had disorder beginning at the age of 35, producing adductor spasm and rigidity of the legs, difficulty in speaking and chronic hyperextension of the proximal phalanx of the great toes with contractions of the peribuccal muscles.

The case is not one of progressive lenticular degeneration or an athetotic syndrome and apparently belongs clearly in no well recognized group.

Progressive Cerebello-Pyramidal Paraplegia Associated with Thermanaesthesia. Syndrome of the Anterolateral Column.—Foix and Valière Vialeix presented a case adequately described by the title. The condition was apparently allied to the Friedreich group but was without a familial history. Syringomyelia was easily ruled out.

Cerebellar Syndrome with Spontaneous Deviation of the Right Index Finger Showing Nothing at Operation.—Moulonguet and Pierre reported the case of a woman, aged 41, who had suffered from a chronic right otitis media since the age of 10. After an attack of vertigo, nystagmus and spontaneous deviation to the right of the right index finger on pointing, an operation was performed to explore the cerebellum. Nothing was found. The patient's condition remained essentially the same. Movements of the head (otogenic origin) and refrigeration of the area over the cranial wound had no effect in changing the symptoms.

Cortical Abscess of Cerebellum Involving Digastric Lobe from Circumferential Fissure to Pons with no Neurologic Signs Except a Terminal Alternating Hemiplegia.—Moulonguet and Pierre reported a case of chronic otitis media in which the patient was admitted to the hospital in a serious condition with fever, severe unilateral headache and vertigo. Sinus thrombosis was found on operation. The patient grew worse, developed aphasia, right facial and left appendicular paralysis and soon died.

Aphasia and Apraxia.—The conclusions of Austregesilo's paper, which upholds Marie's theory of aphasia, are: 1. Aphasia, agnosia and apraxia are usually associated. 2. The tests used by Pierre Marie to demonstrate mental defect also show the coexistence of apraxia. 3. All aphasia patients are apraxic but all apraxic patients are not aphasic. 4. Aphasia appears to be an agnostic apraxia of language and is characterized by the loss of the intellectual powers of speech. 5. The location of the anatomopathologic centers of apraxia is not well known. 6. Clinical observations often reveal symptoms of apraxia in aphasic patients and demonstrate abundantly the principles we maintain, namely, that aphasic and paraphasic patients are cases of true apraxia of speech.

Intermittent Lethargy Indicating the Existence of a Third Ventricle Tumor. Anatomoclinical Report.—Thomas, Jumentié and Chausseblanche reported a case in which the patient had repeated narcoleptic attacks. The first seen by the physicians reporting the case was preceded by headache and vomiting. There was no paralysis. The fundi were normal. Preceding the first attack noted by the physicians there were four, one associated with amnesia and dysarthria, the second without amnesia, the third essentially the same but accompanied by some trouble with the sight, although the fundi were negative, and the fourth was accompanied by headache, vomiting and hiccup. The fundi were negative. The fifth attack showed some diminution of visual acuity, R > L. The patient noted an increase in weight. Periods had ceased prior to the second attack. Polydypsia and polyuria appeared. The fundi suggested retrobulbar neuritis. Three months later there was slight trouble in holding urine and feces. About a month later a sixth attack occurred, accompanied by somnolence, incontinence, a pulse rate of 60, dysphagia, some rigidity of the arms and neck, with a positive Babinski right and left sign and irregular respiration. The patient died eight days later. The duration of the disease

was more than thirty months. The six attacks occurred over a total period of about one year.

At necropsy a large epithelial tumor of ependymal origin, not entirely filling the third ventricle was found. The hypophysis was normal.

The diagnosis was not certain just before death. Epidemic encephalitis was considered. The fundal changes were slight throughout. There was no hemianopsia. The case is interesting on account of the difficulties in making a diagnosis during its long progress.

Rapidly Appearing Blindness with Moderate Papillary Stasis. Death in Coma with Severe, Generalized Contractures. Multiple Areas of Encephalitis Involving the Two Cerebral Hemispheres.—Rochon-Duvignaud, Jumentié and Vallière Vialeix presented an interesting case. The usual sequence, choked disk and blindness, was reversed, the former being slight. The morbid process was not purulent nor like that seen in epidemic encephalitis. The involvement was limited to the white matter, showed large lymphocytic infiltration and neuroglial overgrowth and recalls the "leuko-encephalitis" of Claude and Lhermitte. (*Arch. Neurol. & Psychiat.* **4**:89 [July] 1920.)

With the exception of a mist before the eyes and headache, no symptoms existed prior to the rapid onset of blindness. This occurred in four days. Pupillary reactions to light disappeared. A decompression was done on the right. There was no bulging. Nothing was found. A little less than a month later a convulsive seizure occurred. Then other symptoms appeared—the Babinski sign on the left, incessant twitching of the eyelids, vomiting, headache, then continuous contractions in extension of the legs, in flexion of the arms, rigidity of the neck and to some degree, of the abdomen, and coma. Later the extension of the legs changed to flexion. The patient died seventy-four days after the onset of the disease.

Injections of Air into the Subarachnoid Spaces.—Cestan and Riser have studied certain aspects of the injection of air for ventriculography. The quantity injected should not exceed 20 c.c. and should be between three-quarters and nine-tenths of the quantity of fluid withdrawn. It should be sterile. An aseptic meningeal reaction occurs, the clinical manifestations of which are most often headache, rigidity of the neck and elevation of temperature, none of which last for more than twenty-four hours, and the spinal fluid manifestations of which are polymorphonucleosis and increase of albumin coming on after four hours, reaching a maximum at twelve hours and diminishing in thirty-six hours. On the second day the fluid contains only a few lymphocytes. The albumin may rise to 3 gm. per 1,000 cubic centimeters. The clinical and serologic manifestations are not parallel.

Measurements of spinal pressure with Claude's apparatus showed that the drop resulting from removal of fluid is compensated by the injection of air. In cases showing great increase of intracranial pressure 2 c.c. of fluid are withdrawn at a time and the pressure maintained by injecting the same amount of air.

The air goes into the ventricles when injected into the spinal canal.

Radiological Examination of the Subarachnoid Cavities by the Aid of Lipiodal in a Case of Intramedullary Tumor.—Froment, Japiol and Dechaume reported the injection of lipiodal between the second and third cervical vertebrae in a case clinically suggesting compression of the spinal segments, the seventh and eighth cervical and the first dorsal. The shadow extended down to the second dorsal vertebra but was in a large mass above the first dorsal

vertebra. Then an injection was made into the lumbar region. A roentgenogram was made of the patient head down, and a shadow was found at the sixth dorsal (vertebra). At operation the tumor was located as indicated by the roentgen-ray, from the seventh cervical vertebra to the seventh dorsal. Being intramedullary (glioma at biopsy), it could not be removed.

The authors emphasize the lipiodal injections of Sicard as valuable aids to localization of spinal lesions.

Cyclic Hypoglycemia in Constitutional Psychoses and Particularly in Dementia Praecox.—Dide and Fages have found the blood sugar down to 0.44 to 0.75 (normal is 1.00) in such cases. Spinal fluid sugar was correspondingly decreased. They maintain that this indicates a disorder of the sympathetic system.

Cholesterinemia in Primary Myopathies.—In five case Parhon and Mme. Parhon found the amount of cholesterin had been diminished in two families, one of two, the other of three cases. They recall that the suprarenal cortex plays a part in cholesterin control and also that primary myopathies have been attributed to endocrine disorders.

KRAUS, Paris, France.

DEMENTIA PRAECOX: SOME PRELIMINARY OBSERVATIONS ON BRAINS FROM CAREFULLY SELECTED CASES, AND A CONSIDERATION OF CERTAIN SOURCES OF ERROR. CHARLES B. DUNLAP, Am. J. Psychiat. **3**:403 (Jan.) 1924.

Dunlap carefully studied thirteen brains; eight from cases of dementia praecox from patients whose ages ranged between 20 and 24, and five from controls ranging in age from 17 to 40. The psychotic patients died of bronchopneumonia (2), hemorrhage from ulcer, acute edema of the lungs, decapitation, intestinal and pyloric obstructions, acute bronchitis; two of the control group met violent deaths, one by shooting and the other in a trolley accident, two were poisoned by arsenic and one succumbed to acute peritonitis. The investigation included nerve cell changes, the attempt to identify the acidophil nucleoli staining reaction, lipoid content of nerve cells, neuroglia increase, loss of cells, etc. Dunlap disagrees with the conclusions of Mott, Cotton and others, and is not able to substantiate the hypothetical organic basis for praecox. In the discussion, Orton called attention to the possibility of ultramicroscopic structural alterations, such as are evidently induced by certain acute poisonings, and questioned the reliability of the index furnished by cell counting. He feels that the question of cortex integrity cannot be decided by the number of cells or the cortical measurement, but is dependent on the coefficient of correlation between cell size, cell number and cortex depth.

STRECKER, Philadelphia.

THE DIFFICULTY OF INTELLECTUAL PROCESSES; THEIR PSYCHOLOGY, PSYCHOPATHOLOGY AND IMPORTANCE IN THE INVESTIGATION OF INTELLIGENCE AND DEMENTIA. WALDIMIR ELIASBERG, Schweiz. Arch. f. Neurol. u. Psychiat. **12**:136-144, 201-206, 1923.

A complete investigation of intelligence is not made by a study of aphasia plus apraxia plus agnosia, but is dependent in a large measure on the psychology of association. Residual memories are associated with each other in various combinations. The extent and the relationship of these combinations essentially constitute intelligence. To this might be added attention. The elements of thinking, as considered in the science of logic, do not enter into this.

Heilbronner has called attention to the fact that dementia constitutes something further than the sum total of aphasia, agnosia and apraxia, and that dementia may be added to the foregoing syndrome. Pick has emphasized the fact that we utter thoughts and sentences, not sounds and single words. It is easy to understand why the early studies of intelligence were conducted with meaningless tests, that is, tests which had nothing to do with the accomplishments of intelligence. All that can be said against these methods is that they resulted in nothing. This becomes clear when we note the difference between a so-called test of intelligence and a test devised to demonstrate the ability to perform some practical operation. Occupational tests have been fairly satisfactory. For example, the error made in tests used to determine ability as motorman is only 1 per cent., whereas, the results from the investigation of general intelligence are not nearly so accurate. Eliasberg, in trying to show why this is true states that we can determine the required activities and demands of the particular occupation under consideration, and can imitate these in a test experiment. We are not, therefore, dealing with meaningless tests, but with concrete problems. The general impression that we can supply a good test series only if we understand the underlying psychologic processes, is erroneous. It has been found that the unqualified applicant can be discarded at once on the results of the test, even though there is no knowledge of the underlying psychic processes. It was noted, also, that the candidates fall into groups of the same age, same education and same social milieu. These facts, at first overlooked, assist in making a rough preliminary classification.

The Binet-Simon scale has had the greatest application. It is of value in congenital dementia; however, in acquired feeblemindedness the method is found inadequate, for here loss of memory is not one of the most prominent factors. On the other hand, such factors as attention and imagination are decidedly compromised in acquired feeblemindedness. The feats of memory of a paralytic can be distinguished from those of one with senile dementia according to the type of attention disorder. The Binet-Simon scale tests certain abilities but really does not test intelligence. It is impossible to determine the intellectual age in cases of acquired feeblemindedness. For example, a patient reported by Eliasberg and Feuchtwanger, read like a 6 year old and wrote like a 12 year old; although calculation was limited to numbers below 10 and included only addition, he was completely oriented concerning the prices of bicycles, for he had formerly sold these, and he could count up pieces of money; the marked deficiency of speech impulses scarcely equaled that of a 1 year old child; however, his understanding of speech and his sentence formation corresponded to that of an 8 year old; from a social standpoint, he was still a competent superintendent of a business.

The usual intelligence tests do not take into account sufficiently the emotional setting, such as the submission to the examinatoin and the orientation, both of which are important. It is said that the definition test does not test a new accomplishment, it only tests the clearness of existing information; it is not known how much this is simply a test of aptness in speaking. The indirect, unhistorical, quantitative, comparative methods of measurement must be supplemented by direct, historical, qualitative, comparative methods, that is, by introspective genetic methods. The general impression a teacher has of his pupil is unreliable, as it applies to the undeveloped intelligence of certain groups of persons who are found under definite scholastic influences which do not apply to all forms of intelligence. The introspective method was long mistrusted as

too subjective; however, its value and practical importance need no longer be questioned and defended.

The encountering of difficulties are fundamental experiences in our intellectual lives. Whether we approach a problem with energy and confidence or with neurasthenic insufficiency determines whether or not the problem will be solved. In discussing the disturbances of speech, Pick noted that the greatest disturbance occurred in expressions that are least habituated, least automatic and most under the influence of conscious direction. Conscious direction is required especially in the finding of words; grammatical processes, on the other hand, are more nearly automatic. The greatest difficulties are experienced in the agrammatic formulation of the telegram style, in the acquisition of dialects, in slowing of the speech tempo, in changes of rhythm, in the search for words, in diversion of attention, in a knowledge of the difficulty of a task, in complete surrendering to the task and in affective disturbances of an unpleasant nature. A problem is considered as difficult objectively when it is unusual and when it can be accomplished by few or no persons.

In order to demonstrate the difficulty of intellectual processes by the method of self observation, adult, educated subjects were chosen for two reasons: first, to secure uniformity of intelligence so that tests which these subjects found difficult could undoubtedly be so designated; and second, so that the self observation of these subjects would give us some understanding of the phenomenology of the difficulty. In mentally well developed persons, mental processes can be made difficult by disturbing them. Such a disturbance can be produced by simple interruption, as advocated by Baade. The thinking process may be fractionated by directing attention to different matters at different intervals, as Henning suggested. Pauli, in using the Masselon test, in which sentences are formed from several words, presented these for a given length of time on a card, and while the subject was occupied with this problem, two tactile stimuli were given with instructions to determine which of the two was the harder. The application of the tactile stimuli was not rhythmic, and the intervals were varied constantly. Furthermore, the subject was not told what type of examination or combinations of examinations would be required of him.

Among the results that can be recorded objectively, and this is also obvious to the subject, is the length of time required for the solution of the problem. It is rather characteristic for the subject to deny responsibility for the solution of the problem by saying that nothing occurred to him. Characteristic expressions of effort, such as clearing the throat, flushing of the face and wrinkling of the forehead are noted. When a difficult problem is presented the subject usually asks himself, "What is the meaning of this test?" In the solution of difficult problems there is a wide fluctuation of attention and in the level of consciousness. This is not so marked when simple problems are submitted. An important criterion in determining the question of difficulty is the condition of the affect. The results of this method are inaccurate as they demand a comparison of degrees of pleasure and displeasure; if we are content, however, with results not mathematically exact, a great deal of value remains. Another criterion is the evaluation of a finished piece of work. Thus, the subject may be dissatisfied with the solution, may be uncertain of the result; other solutions may occur to him that are better. The condition reminds one of so-called "step-up" jokes. Psychologically, the degree of difficulty can be estimated only by knowing the entire psychic context of the given period. The estimation of difficulty also rests on past experience. Another aid in estimating the degree of difficulty may be had if we know that the subject used some

method of help other than the one demanded for the solution of the problem. For example, when asked to subtract 19 from 38 it may occur to him that 38 equals 2 times 19, which is a much simpler operation than that of subtraction, and yet gives him the solution. It is only by a complete protocol of the mental activities carried on by the subject that such things become evident.

All thinking requires the will to think. It has been seen that the criteria used in evaluating the degree of difficulty of solving a problem are concerned with volition and feeling. It is therefore necessary to have determined previously that the intelligent subject is not deficient in willing and in feeling. A neurasthenic subject with the feeling of insufficiency, although intelligent, could not be used for these examinations. It is well known that where there is a persistent or transient inability to solve a problem, perseveration arises. Many sick persons do not judge perseveration correctly and do not regard it as an evidence of difficulty, even though this thought may occur to them. The very awareness of difficulty implies a certain degree of intelligence. The difficulty of judging intelligence is particularly great in cases of sensory and motor aphasia combined with apraxia and agnosia. In these cases, the betrayal by a slight gesture that the patient recognized his deficiency is an extraordinarily accurate and safe criterion of intelligence. Socrates seeks the criterion of intelligence, not in the outstanding accomplishment, but in the attitude of the person toward his own accomplishment. From this can be learned the limits of their ability. He says, "I noted at the same time that their gift for poetry misled them into the belief that their wisdom transcended the wisdom of all others in all other things of which they understood nothing. Their works are not the fruit of wisdom but of a natural inclination and enthusiasm such as is seen in fortune tellers and singers of oracles." (Apology.)

The attitude of persons toward their abilities has been investigated a number of times. Anton noted that in serious defects, such as complete blindness and deafness, the appreciation of this defect may not occur and the person may maintain that he can see or hear. In Anton's cases these defects were due to central lesions, which he thought furnished sufficient explanation for this striking and peculiar lack of insight. Redlich and Bonvicini, however, described cases in which the blindness was peripheral in origin. In one case the blind person could be convinced of his blindness only by having his environment described with great accuracy; this description was quite different from the mental picture that the blind person had had concerning his environment. Stertz does not believe that this symptom represents an intelligence defect, as blindness is usually noted by markedly mentally deficient persons who are blind. He believes that it is comparable to the psychologic mechanism of displacement. Eliasberg thinks, however, that it is only on the basis of defective intelligence that such a psychic mechanism can take place in the face of such a marked defect as blindness.

In concluding, Eliasberg defines intelligence as an active attitude of the personality toward his environment. Dementia is a deficiency of intellectual accomplishment. The question arises whether dementia can disturb the personality. We must decide, if we can, whether there is an insight in regard to the defect; if this is present, we must not speak of dementia. Where primitive functions occur that merely represent a passive adjustment to a situation from a biologic point of view, one may get the impression that personality directed toward a given end is maintained. In this case, however, we must speak of a dementia personality.

Concerning how this hierarchy of points of view that constitutes intelligence is developed, we know little. Besides the investigation of psychologic processes in children, an investigation into the mechanism of thought in primitive peoples and in pathologic cases might be utilized.

For practical purposes, the foregoing methods of investigating intelligence supercede the prevalent practice of attempting to evaluate intelligence with numerical accuracy. Certainly the objection cannot be made to the method that it is not intelligence that we are investigating. This method applied to the investigation of dementia represents a new direction. Up to this time, the frequent difference of opinion between the results of the usual psychometric determination and the subjective impression of the psychiatrist was always decided in favor of the latter. This unsatisfactory state of affairs can be remedied only by the utilization of better and more scientific methods.

HENRY WOLTMAN, Rochester, Minn.

FURTHER CONTRIBUTIONS TO THE STUDY OF THE EVOLUTION OF THE FOREBRAIN. J. B. JOHNSTON, Parts I to IV, J. Comp. Neurol. **35**:337, 1923; Part, V, J. Comp. Neurol. **36**:143, 1923.

This paper deals at length with the problem of the development and differentiation of the basal surface of the brain and of the temporal pole particularly. Some reflections on the evolution and form changes of the forebrain are incorporated in the fifth part.

The Corpus Striatum.—In the human embryo of 18 mm. from the ventrolateral wall of the forebrain vesicle, that sector assigned by Herrick to the corpus striatum, two ridges project into the lateral ventricle. They are separated by a deep, transient, ventricular groove, the sulcus striocaudatus, which is a permanent structure in some reptiles. The dorsal ridge becomes in mammals the chief part of the tail of the caudate nucleus. It is thought to be formed by an infolding of the lateral margin of the general pallium along the line of the rhinal fissure. The ventral ridge becomes the bed nucleus of the stria terminalis. Anteriorly it is continuous with a mass of cells lying in the head of the caudate at the ventral angle of the ventricle and covered by the cortex of the tuberculum olfactorium. As this area, Johnston's nucleus olfactorius anterior, is traced caudally, it becomes divided into two limbs by a ventricular groove which is the continuation of the sulcus striocaudatus. The medial limb is the nucleus accumbens, a part of the corpus striatum, not to be confused with the septum or area parolfactoria; on the contrary, it must be recognized as a secondary olfactory center and as such distinguished from the rest of the striatum and the parolfactory nuclei. The lateral limb forms a large portion of the head of the caudate nucleus, which portion is, therefore, also a secondary olfactory center continuous rostrally with the nucleus olfactorius anterior and caudally forming a band of gray matter in the medial border of the nucleus caudatus and forming the bed of the stria terminalis and anterior commissure. The bed of the anterior commissure is formed by a proliferation of the cells of the bed of the stria and a migration toward the midplane to form a bridge through which the fibers may afterward cross. The bed of the stria remains distinct back to the amygdala, where it expands and the fibers of the stria distribute to the various amygdaloid nuclei. The caudate nucleus is, therefore, formed by a fusion of two distinct cell masses: (1) ventrally and medially the bed nucleus of the stria which has olfactory connections, and (2) a dorsal and lateral component with no olfactory con-

nections, the caudate proper. In the following description these are called respectively the stria bed and the caudate nucleus, although both are included in the caudate of the classical nomenclature.

Although the putamen arises similiarly to the caudate nucleus from the lateral part of the primary general pallium, it seems to be formed earlier in ontogeny than is the ridge which becomes the caudate nucleus. Again, there is evidence that the putamen and the globus pallidus are present in fishes as a derivative of the somatic area, while the caudate nucleus appears for the first time in phylogeny in reptiles. There is less distinction between the caudate and the putamen in lower mammals than there is in man. Their cells are of similar type, and they are continuous between the fibers of the internal capsule. In all mammals they fuse behind the caudal border of the internal capsule and become lost among the nuclei of the amygdaloid complex. Since both the putamen and the nucleus caudatus establish their functional connections through the internal capsule and resemble each other in histologic structure, they are probably closely related in function; and by virtue of their origin from the general pallium they are somatic mechanisms.

The globus pallidus can be readily distinguished from the putamen by the presence of large cells, especially along the inner margin of the globus next to the putamen, and by its proximity to the internal capsule. This nucleus can be followed from the caudal border of the internal capsule where it lies in contact with the central nucleus of the amygdala forward to the anterior commissure where it lies medial to the lower border of the putamen.

The corpus striatum consisted primitively of an olfactory and a somatic component, the bed of the stria terminalis and the lentiform nucleus, respectively. To this has been added later in phylogeny a derivative of the general pallium, the nucleus caudatus, with functions probably of a similar but somewhat lower order than those of the neopallium. The nucleus caudatus has become associated functionally and morphologically with the nucleus lentiformis, while the bed of the stria terminalis has retained its primitive connections with the olfactory centers and its anterior and posterior parts have become prominent olfactory nuclei.

The Secondary Olfactory Centers.—The development of these in the selachians is described and compared with their development in reptiles and mammals. The medial olfactory area of the selachians gives rise to the medial parolfactory and the anterior olfactory nuclei of higher forms. Subsequent to the elongation of the hemisphere in reptiles and mammals the lateral olfactory area was carried into the temporal pole and has become the pyriform lobe and part of the amygdala. A connection is retained between the amygdala and the medial olfactory area in front of the anterior commissure through the diagonal band. At this level the medial and lateral olfactory areas are joined along the ventral surface of the hemisphere by the basal olfactory area. The rostral part of this basal area forms the tuberculum olfactorium of reptiles and mammals, while the caudal portion probably becomes the gray matter of the anterior perforate space and diagonal band. The diagonal band consists of a diffuse system of fibers, some of which connect the amygdala with the hippocampal formation, while others connect the former with the tuberculum olfactorium.

The Amygdaloid Complex.—One of the significant features of this paper is the detailed analysis of the stria terminalis and the amygdaloid complex. In marsupials and higher mammals Johnston recognizes five principle bundles within the stria terminalis: the commissural, hypothalamic, infracommissural

and supracommissural and the stria medullaris bundles. The amygdaloid complex consists of six principle nuclei which may be divided into two groups: namely, the more primitive central, medial, cortical, and the nucleus of the lateral olfactory tract; and the newer basal and lateral nuclei.

The nucleus of the lateral olfactory tract is a remnant of the primitive lateral olfactory area of fishes and has remained as a terminus for some of the fibers of the lateral olfactory tract. It is connected through the anterior commissure with its fellow of the opposite side by the commissural bundle of the stria terminalis. Also the stria medullaris bundle connects this nucleus, the tuberculum olfactorium, the hippocampus, and the pyriform cortex with the habenula. The central and medial nuclei, which have also been derived from the lateral olfactory area of fishes, always remain in close relationship and together give rise to the hypothalamic bundle of the stria terminalis which connects these nuclei with the hypothalamus. This tract is homologous to the olfactory projection tract of Cajal. It lies above and medial to the internal capsule. The medial nucleus maintains a superficial position next to the caudal end of the hippocampus, while the central nucleus is found beneath the lentiform nucleus and internal capsule. The connections of the central and medial nuclei indicate that they are concerned with olfactogustatory correlations. The cortical nucleus is derived from a part of the primitive olfactory area caudal to the central and medial nuclei.

The lateral olfactory area of selachians is marked off from the basal area by a groove, the sulcus endorhinalis. With the caudal elongation of the hemisphere, this sulcus comes to lie within the pyriform area and parallel to the rhinal fissure. Caudally this groove becomes the site of proliferation and immigration of cells and infolding of the brain wall. This part of the sulcus is called the fissura amygdalae. The lateral and basal amygdaloid nuclei have been formed from the anterior and posterior parts of this infolding, respectively. The basal nucleus contains a large and a small cell component. In the opossum an accessory basal nucleus is also present. The basal nuclei are connected above and below the anterior commissure with the parolfactory area by means of the supracommissural and infracommissural bundles of the stria terminalis. There are also associational connections with the pyriform cortex. The lateral nucleus is covered over its whole lateral surface by the external capsule fibers and is more closely related to the putamen than any of the other amygdaloid nuclei. The chief connections of the lateral nucleus are through associational fibers with the external capsule, the putamen, pyriform cortex, and the diagonal band. It is, then, an olfactosomatic correlation center. These newer nuclei appear to have resulted from expansion and further differentiation within the pyriform lobe, which process may be associated with an increased demand for olfactosomatic correlations dependent on the assumption of a terrestrial habitat with its new and varied tactile, visual, auditory and proprioceptive stimuli.

That part of Johnston's description of the stria terminalis and the amygdaloid complex which deals with the condition existing in the opossum has been carefully checked by Professor C. Judson Herrick and has been found to be exceedingly accurate and objective.

The General Plan of the Forebrain.—Kingsbury has recently introduced a modification of His' interpretation of the relationships existing in the anterior part of the neural tube. This author departs from His' classical conception that the notochord extends forward, accompanied by the parallel basal and alar plates, as far as the primitive infundibulum and claims that the notochord

extends only to the fovea isthmi and that cephalad to the notochord the basal and alar plates of either side are continuous across the midplane. Caudal to the fovea isthmi the fusion of the basal plates is marked by a median seam, the neurochordal plate which represents the line of closure of the blastopore and which persists in the adult as the median raphé. The brain may be divided into an anterior, nonmetameric, prechordal brain which posseses no true raphé and a more caudal metameric, epichordal brain which does. The nonmetameric brain contains no motor mechanisms, the basal plates of His do not extend into it, the sulcus limitans cannot be recognized in it, and the floor of the neural tube in this region is formed by the alar plates of the two sides which are continuous across the midplane. The alar plates here give rise to the dorsal portion of the midbrain and the entire forebrain (telencephalon and diencephalon). In front of the fovea isthmi in the region where the basal plates are continuous across the midplane lie the median and lateral nuclei of the oculomotor nerve. This portion of the midbrain should be regarded as a transitional zone between the nonmetameric and the metameric regions, the latter beginning with the cerebellar segment. This region presents some metamerism in the possession of (1) motor nerves (third), and (2) the neurochordal plate in younger and raphé fibers in older stages. It represents the extreme anterior angle of the fused lips of the blastopore. Behind the fovea isthmi the epichordal brain shows true metamerism. Johnston apparently accepts Kingsbury's conclusions with some modification.

Form Changes in the Evolution of the Hemispheres.—Some factors which operated in the localization of function within the premetameric region are reviewed, such as: (1) the presence of a visual organ developing within the somatic sensory column; (2) the olfactory and gustatory organs stimulating the development of a special visceral column; and (3) the fiber systems ascending from the metameric brain. Among the last there may be mentioned especially the general cutaneous and proprioceptive systems which enter the dorsal part of the thalamus and the gustatory and general visceral tracts which enter the hypothalamus. Although in the lower fishes much of the telencephalon is dominated by olfactory impulses, Johnston believes that a general pallium exists independent of olfactory centers. This view is based on (1) the ascending tertiary systems from the tactual and visual fields of the thalamus (thalamic radiations), (2) the tractus pallii which ascends from the hypothalamus carrying gustatory and other visceral impulses into those olfactory centers from which the hippocampus is formed in higher vertebrates; and (3) the possession of a pallial commissure (corpus callosum) independent of the hippocampal commissure. Within this somatic area correlations are effected with tactual, visual, thermic, painful, gustatory and proprioceptive impulses. These new integrations find their efferent channel through the connections with the anlagen of the corpus striatum. This view departs radically from that of Elliot Smith and Herrick who have pictured the general pallium as developing within olfactory centers and gradually becoming emancipated from them during evolution, leaving a neopallium for the integration of purely nonolfactory impulses and an archipallium (pyriform lobe and amygdaloid complex) for olfactosomatic correlations.

The phylogenetic history of brain structures has been that of a continuous modification of old elements inherent in our lowly ancestors under the influence of changing environment and habits; no fundamentally new structures have appeared. This is well illustrated in the development of the somatic area or neopallium. As described above, Johnston recognizes the presence of a

primordium neopallii in selachians. Reptiles are accredited with a differentiated neopallium with beginning localization of function as shown by the motor response to electrical stimulation. The growth of the neopallium became the dominating influence in the rearrangement of parts within the forebrain. The development of the pyriform lobe and amygdaloid complex in association with the increasing importance of the thalamic radiations has already been described. The lateral expansion of the neopallium caudal to the olfactory peduncle and the reduction of the medial olfactory nucleus rostral to the peduncle (as in selachians) were influential in the migration of the olfactory bulb from a lateral to a rostral and median position. Further expansion of the pallium produced a lateral bend in the crus cerebri and lentiform nucleus. The medial walls of the hemisphere approximated each other and the corpus striatum came to occupy the floor of the lateral ventricle. Further changes in the striatum were described above. As the pallium extended caudad the pyriform lobe came to cover the lateral surface of the corpus striatum. Finally an insula was formed by projecting opercula of neopallium. When the hemisphere bent down to form the temporal pole the structures covering the ventricular surface of the lentiform nucleus and crus cerebri were carried into this pole. Finally, because of the enormous development of a frontal lobe rostral to the motor area, the olfactory bulb came to lie upon the basal surface of the frontal lobe.

GRAY, Chicago.

THE SYNDROME OF THE LONG RADICULAR FIBERS OF THE POSTERIOR CORD. JUMENTIÉ, Rev. neurol. **29**:432 (April) 1922.

This second paper, by Jumentié, is on a subject suggested or inspired by Déjerine when Jumentié was a student under Déjerine. The latter styled it the syndrome of the long radicular fibers of the posterior cord. Its two outstanding features are integrity or preservation of superficial (tactile) sensibility, and loss, even to complete destruction, of deep sensibility. It is, in other words, a dissociation syndrome. It is like the tabetic dissociation syndrome in that deep sensibility is affected, but unlike it in the preservation of superficial sensibility. The author then refers to cases described by himself in conjunction with Déjerine (1914); in a case presented by Déjerine and Mouzon (1914); and in a case described by Mme. Déjerine and Mouzon (1915).

The author's paper is based on the anatomopathologic examination of two cases. They are cases designated by the author as subacute combined sclerosis and cord changes mentioned for the first time by Lichtheim (1887) when he was discussing pernicious anemia. Lichtheim clearly differentiated these pareto-ataxic states from tabes, which they resemble in certain respects. Déjerine clearly established this difference, clinically in contrasting the syndrome ot the long radicular fibers of the posterior columns of the cord of the tabetic type of sensory dissociation with tabes complicated by sclerosis of the lateral columns. He also showed the topographic differences of the two varieties of combined sclerosis.

Lichtheim, then Minnich (1892), pointed out the following important anatomic aspects: (1) the integrity of the posterior roots and the topography of the lesions in the posterior columns, especially the outer part of the column of Burdach; (2) two types of medullary lesions. Then Thompson (1911-1912) showed the almost complete preservation of superficial sensibility and the marked loss of the sensation of passive position and disturbances of tactile discrimination. He showed that the lesion was limited to the posterior columns.

In spite of his own proofs and facts, he regarded this syndrome as a tabetic type of disorder.

Jumentié then gives in detail a case report, both clinical, and anatomopathologic, of a man, aged 48, affected with flaccid paraplegia which had occurred eight days before he entered the hospital. The paraplegia had been preceded a year before by spasticity of the lower extremities, and accompanied by general asthenia, but no definite anemia. In the hospital he showed loss of the tendon reflexes, a double Babinski sign, ataxia of the upper extremities and vesical retention. He also suffered from severe pains following both passive and active movements. There was also a definite loss of deep sensibility, but a preservation of superficial sensibility (touch, pain and temperature); also loss of sense of motion and position up to the hips.

Postmortem, laboratory examination revealed:

1. Old sclerotic lesions, degenerative in character, involving the column of Goll and the inner portion of the column of Burdach.

2. Degenerative lesions, sclerotic, less pronounced, and of unequal intensity, of the pyramidal tracts.

3. Swelling of myelinated fibers in the zones of invasion most recently affected and likewise in the uninjured portions; swelling of the cerebellar tracts and the direct pyramidal tract.

4. Striking preservation of posterior roots at the point of their penetration of the posterior gray matter.

5. Pseudosystematized aspect of sclerosis of the columns of Goll and Burdach.

The histologic picture is that described by Mme. Déjerine and André-Thomas in their essay on lesions of the cord in anemia (1899).

The vessels in the sclerotic areas have thickened vessels; in the invaded areas, cribriform in appearance; the dilated capillaries show a marked diapedesis and an encroachment of granular bodies.

The second case was that of a woman, aged 49, with a profound anemia. Her gait was hesitating in character, somewhat incoordinated, with lateral oscillations and occasional falling due to vertigo, these disturbances being aggravated by occlusion of the lids. There was no trace of paraplegia. The tendon reflexes were normal; there was no Babinski sign. Sensibility to touch, pain temperature and deep pressure was almost intact. Vibration sense (on bony prominences), sense of position, discrimination of two points—all were *distinctly* impaired. There was marked astereognosis. Sphincter control was good. The patient died after sixteen hours of thoracic pain. The duration of the illness was eight and one half months.

The histologic lesions were the same in this case as in the preceding. A comparative study of the two cases showed lesions in the second case sharply limited to the posterior columns, but more severe and extended in the second.

The degree of intensity and duration of the destructive processes on the nerve fibers determines the degree of anatomic changes. Discrete lesions in the posterior columns may give rise to no perceptible symptoms during life, and are usually noted only after necropsy. Studies of various investigators bring out the following types: (1) a light or mild paretospastic state with slight ataxia and paresthesia; (2) severe spastic paraplegia with anesthesia of the trunk and legs; (3) complete flaccid paraplegia with absolute anesthesia. There may also occur paralysis of the sphincters and edema.

Demyelinization and destruction of the axis cylinders are usually limited to the columns of Goll and Burdach, that is, to the long radicular fibers which conduct deep sensation.

If we compare these lesions, so sharply localized, with those of tabes complicated with paraplegia, we see that in the tabetic scleroses, the lesions affect the posterior roots, involving all the fibers without distinction, thus modifying tactile sensibility as well as that of heat and pain.

From the foregoing, it is evident that the term "syndrome of the long radicular fibers of the posterior cord" is not synonymous with "combined subacute sclerosis of the cord," since the first term is related only to the sensitive stage, the first, in point of time, of this disorder. This symptomatologic concept is of the utmost importance as it enables us to make a delicate differential diagnosis at the very beginning, especially when anemia, so often an associated symptom, might lead us astray in determining secondary manifestations.

It is probable that since the attention of observers has been drawn to this syndrome, deep sensibility being studied with greater care may appear to show itself disturbed with greater frequency.

It should be noted that this symptomatology is found in other conditions. Déjerine and André-Thomas report it in pernicious anemia, ergotism and pellagra; it might also appear at certain stages in the evolution of a cord tumor or a gumma, or it might appear in certain forms of hematomyelia. Moreover, by the vascular route, an infectious organism may produce similar symptoms. Alquier (1909) reported this in a syphilitic patient ten years after the specific infection.

The author then presents his arguments, in spite of contrary opinion, for the syndrome he is describing relative to the part the vascular supply of white matter plays in such disorders. He considers them true vascular lesions, i. e., thickening of the arterial walls, not so much the cause of the sclerosed fibers or columns, but rather as the end result.

The syndrome described by Déjerine, to which the present study is dedicated, is above everything else a syndrome of localization. It corresponds to a special topographic lesion; namely, destruction of long root fibers of the posterior column which conduct deep sensibility, sense of position, vibration sense, stereognosis, to the exclusion of the more central short fibers which are the pathways for superficial sensibility such as touch, pain, and temperature.

JONES, Detroit.

INTERVERTEBRAL COMPRESSION OF THE SPINAL NERVES. EDITORIAL, J. A. M. A. **82**:34 (Jan. 5) 1924.

It has sometimes, unjustly, been alleged that physicians have paid too little attention to compression of the spinal nerves in the intervertebral canals and foramina as a cause of symptoms. Probably the most fruitful studies of this difficult subject have been made by French investigators who have outlined types of vertebral disease and the syndromes connected with them. The investigations of the intervertebral canals in the living subject, even with the aid of the roentgen rays, is extremely difficult, owing to the complexity of bony processes in their vicinity and the variation in the direction of the canals at different levels of the spinal column. The interpretation of roentgenograms is correspondingly liable to error and has proved of little practical value. There has also been a tendency on the part of neurologists to explain all symptoms

that indicate affection of spinal roots by involvement of the roots in inflammation of the dura.

Sicard, in particular, has done much to throw light on this obscure problem, and some time ago described a condition of inflammation of that portion of the spinal root that lies within the intervertebral canal under the name funiculitis. This was distinguished from radiculitis, which is an affection of the intradural portion of the root that lies between the spinal cord and the intervertebral canal, and also from involvement of the trunk of the nerve root that extends from the foramen to the nerve plexus. The funicular portion of the root, lying within the channel between the vertebrae, is outside the dura mater and is embedded in a mass of "adiposocellular" tissue, which is continuous with the fatty layer that surrounds the dura beneath the bones and ligaments, and constitutes the so-called epidural space. In 1901, Sicard and Cathelin introduced the method of epidural injection of cocain in the treatment of intractable sciatica and other pains, entering this space through the sacrococcygeal joint. Recently, Forestier,[1] a pupil of Sicard, has published a monograph in which are detailed the results of extensive research, both anatomic and clinical, on the funicular portion of the spinal nerves. He describes a technic by means of which he has succeeded in injecting the epidural space by inserting the needle between laminae in the lumbar and dorsal regions. Using a radiopaque solution of iodin in a vegetable oil, which is said to produce no tissue reaction and to be entirely harmless, he has succeeded in securing roentgenograms that demonstrate the dispersion of the injected fluid through the epidural space and thus the presence or absence of obstruction in the space. Obstruction had been observed only in one case of Pott's disease and another of medullary tumor; in cases of chronic "rheumatic" affection and various forms of spondylitis and spondylosis, the injected fluid spread freely. Since then, Sicard and Forestier have recorded[2] the finding of obstruction in numerous cases of tuberculous and carcinomatous disease of the vertebrae.

Swelling of the epidural adiposocellular tissue, however, has been observed directly at operations, and its presence has been deduced from the condition of the spinal fluid, in patients with chronic lumbago and sciatica. As was pointed out by Sicard and Foix in 1910, the spinal fluid in funiculitis contains an increased amount of albumin without pleocytosis. The dissociation distinguishes the fluid from that in meningitis, in which both the albumin and the cell content are increased. The increase of albumin in funiculitis is explained as the result of interference with fluid circulation from compression of the venous and lymphatic vessels in the intervertebral canals.

Some of the conclusions reached by Forestier are of particular interest in connection with the much exploited intervertebral compression of spinal nerves. He points out that compression of the nerves and other structures within the canal gives rise to a definite syndrome: local pain, spontaneous or provoked by movements of the vertebral joints, and accompanied by visible and palpable rigidity of the spinal muscles, with limitation of movement and altered posture; neuralgic pain radiating into peripheral parts that is influenced by movements of the trunk and limbs and also by coughing; objective sensory disturbances that are often absent, and consist rarely of hyperesthesia, more

1. Forestier, Jacques: Le trou de conjugaison vertébral et l'espace épidural, Paris, Jouve & Co., 1922.

2. Sicard and Forestier: L'huile iodée en clinique, Bull. et mém. Soc. méd. d. hôp. de Paris **42**:309 (Feb. 23) 1923.

commonly of hypesthesia to pin-pricks in the area of skin corresponding to the root affected; rarely motor symptoms, which are always slight and include diminution of power and clumsiness of movement; alteration of tendon reflexes, most often, in the lower extremities, in the direction of exaggeration, less commonly diminution. Vasomotor and pilomotor disturbances have not as yet been observed. To these must be added the changes in the spinal fluid already described.

Perhaps even more significant is the conclusion reached concerning the effects of movements of the vertebrae on the size of the intervertebral canals. Sliding of the articular surfaces on one another results in an actual enlargement of the canal and it is not until the upper articular process passes over the summit of the lower one, that is to say, until complete dislocation occurs, that compression of the contents of the canal will be produced. Furthermore, such displacement will at once "crush the contents of the intervertebral canal, which becomes *reduced to a merely potential space*." The consequences must inevitably be complete interruption of nerve function with loss of sensation and paralysis of muscles corresponding to the distribution of the root involved.

These observations, which are incidental to the purpose of Forestier's investigations, establish clearly the conditions under which compression of the spinal nerves will occur and the symptoms that follow from such compression. They leave no room for speculation, exaggeration or exploitation.

ARM-CHEST ADHESIONS: BRACHIOTHORACIC ADHESIONS, AXILLARY WEBS. JOHN STAIGE DAVIS, Arch. Surg. **8**:1 (Jan.) 1924.

Davis reports the results of the use of surgery in the treatment of brachiothoracic adhesions or axillary webs. Forty-eight cases form the basis for this paper. Burns with fire were the cause of the condition in forty-four cases, with hot grease in one case, roentgen-ray burns in two cases and slough of a breast flap in one case. Twenty-eight cases occurred in children below the age of 10; ten other cases in the second decade. The cases were seen in less than two years following injury in thirty patients, up to ten years in thirteen. Forty-one patients were operated on. The types of operative procedures are grouped in a general way as: (1) excision of binding scar with suture, four; (2) transverse division with vertical closure, three; (3) division of excision of binding scar with autoplastic operation, thirty-four. Secondary operations numbered eighteen. The results were: well, twenty-five; improved, sixteen.

The extent of the deformity varied with the causative agent. Burns by fire were the most serious, producing more or less severe scarring in addition to the arm-chest adhesions. The axillary webs showed marked differences in type from the thin lax membrane which allowed considerable motion to the solid thick mass of scar tissue which obliterates the armpit and glues the arm immovably to the chest well; these adhesions may vary in extent from a few centimeters to the full length of the arm. The entire axilla may be obliterated, or the adhesions may be along the anterior or posterior axillary fold, or both. The amount of motion possible for the arm and shoulder depends on the vertical length of the web and on its shortness transversely. The greater the vertical length and the shorter the transverse measurement, the less the motion. The anatomy of the axilla is often obliterated by the scar tissue making extensive dissection difficult and dangerous.

In cases of long duration frequently marked atrophy of the adjacent bones (humerus and parts of the scapula) and also of the muscles and other soft parts appears, due to disuse, with the additional factor of pressure from the

thick unyielding scar. The atrophied arm may be thinner, the humerus may be shorter, the head of the humerus may be pulled forward by the scar; contractions of muscles and muscle aponeuroses may be noted. Every case with extensive adhesions showed a scoliosis which could be corrected only by release of adhesions and relieving scar tension. Chest expansion is often interfered with on the affected side, and rib deformities are seen at times.

Prior to operation every effort should be made to build up the patient's nutrition and general health. Great care must be taken to make the dressing of the wounds as painless as possible, for nothing so breaks down a child's morale as the dread of a painful dressing. Postoperatively, the attention should be focused on making the patient as comfortable as possible and at the same time preventing the recurrence of contractures. The wound should be inspected frequently and any tension on flaps relieved or tendency to infection combated. Skin grafting is often necessary. After healing is complete, passive movement, massage and stretching exercises are of great value in obtaining the full benefit of the operative procedure.

THE SURGERY OF THE PITUITARY BODY WITH ILLUSTRATIVE CASES. CHARLES H. FRAZIER, Arch. Surg. **8**:39 (Jan.) 1924.

Frazier discusses the treatment of primary intrasellar lesions. At present these conditions may be dealt with apparently most satisfactorily by the transsphenoidal approach. He quotes eighteen consecutive cases from his service, without a death.

Conclusive evidence in the diagnosis of pituitary lesions usually is revealed in the roentgenogram. The dimensions of a normal sella should not exceed 10 by 12 mm. The visual fields in primary intrasellar lesions are well known. The vast majority of patients exhibit a bitemporal hemianopsia. The vision usually begins to fail in one eye first. Many patients are blind in one eye before they appeal to the surgeon for relief. Headache and endocrine disturbances, acromegaly or the Fröhlich syndrome are the other symptoms commonly seen in this condition. When other cranial nerves beside the second are involved, and neighborhood symptoms and evidences of intracranial pressure appear, the lesion may be presumed to have reached such a size as to have escaped the confines of the sella and become an extrasellar as well as an intrasellar growth.

Frazier has had no success with glandular therapy in the treatment of pituitary disease. Radiation and roentgen-ray therapy has been of value in certain cases. But for the relief of headache of pituitary origin and to restore vision, lost through pressure of the growth on the optic nerves and chiasm, sella decompression by the sphenoidal route has proved satisfactory and is the method of choice in dealing with primary intrasellar lesions.

GRANT, Philadelphia.

HEREDITY AND OBESITY. EDITORIAL, J. A. M. A. **82**:470 (Feb. 9) 1924.

If a hundred men of about the same stature are compared, Davenport[1] remarks in an unusually elaborate study of body build and its inheritance, it is seen that they vary greatly in weight. At the same time they vary in form, and especially in bulk. This variation is popularly recognized by the variety

1. Davenport, C. B.: Body-Build and Its Inheritance, Publication 329, Carnegie Institution of Washington, December, 1923.

of terms applied to build. The slim, gaunt and lanky, the obese, chubby and portly—these and many more designations have been adopted to describe persons of unlike "build." The extremes of such structural make-up of man not infrequently come to the physician for consideration in his professional capacity. He is expected to transform the slender and the corpulent, as the case may be, into a more nearly average type of body build; to eliminate the supposed disadvantages of undersize and the alleged dangers of overweight by the promotion of more nearly ideal form and weight.

Were the features just stressed merely the expression of an inappropriate adjustment between intake and output of matter, it would seem that the correction of objectionable build ought to become a comparatively easy task. Gain or loss would call primarily for more or less favorable nutritive balances regulated by dietary control. There are many, indeed, who look on obesity as essentially the expression of undue food intake. Others regard the condition as one involving in some way a disordered metabolism. It has even been suggested that the obese digest and utilize their food more efficiently than persons belonging to the nonfattening group, thereby putting on weight under dietary conditions that fail to promote gains in most persons. Those who reject any peculiarity of metabolism in the fattening process point to certain habits of life as favoring obesity. Thus, to quote a recent writer, Eskimos are assumed to grow fat because they eat blubber and huddle in narrow spaces, undergoing little movement throughout the long, dark winters.

It is doubtless true that the ability of the body to store proteins and carbohydrates is limited so that any considerable and long continued excess of food leads to an accumulation of fat. Obesity arises only when the intake of energy in the food has exceeded the expenditure. There is no extensive experimental justification for assuming that the basal metabolism or fundamental energy overturn of obese persons is lower than the lowest normal limits. Nevertheless, it is not easy to avoid the conclusion from every-day observation that certain persons grow fat despite the fact that they appear to eat moderately and appear to take an ordinary amount of exercise. Whatever the fundamental cause may be, says Davenport, the fact remains that in certain families there is a widespread inclination to the production of slender individuals, while in other fraternities certain proportions (though usually not all of any fraternity) are fleshy or even obese. Perhaps, as in the case of the Jersey as contrasted with the beef steer, the two kinds of individuals do not metabolize their food in the same way; some are spare and muscular, others lay on fat. In any case, we cannot disregard the constitutional factors in build.

From his investigations of heredity as expressed in family studies of body build, Davenport can see that no other theory than that constitutional differences as well as nutritional differences determine build is sufficient to meet all the facts. From the standpoint of the geneticist's evidence, variations in build are to be accounted for not merely by variations in intake and outgo of calories, but also by the endogenous factors that determine the "economy of nutrition" or the cost in energy of adding an additional kilogram of weight to the body. The factors involved in producing differences in these respects are hereditary factors. The hereditary factors probably work through the participation of special organs that influence metabolism, notably the endocrine glands. The latter thus intermediate between the chromosomal constitution, on the one hand, and control of metabolic processes, on the other. One naturally thinks of the influences associated with the thyroids and the pituitary glands.

That other constitutional conditions than those of the larger endocrine glands may play an important part in metabolism, says Davenport, cannot be denied; probably the quality of the protoplasm of every active cell influences the bodily metabolism; but the endocrine glands proper seem, as it were, to be told off for this specific purpose, and thus peculiarities in their functioning lead to striking results.

From the statistics collected by the Eugenics Record Office of the Carnegie Institution of Washington, it appears that the diseases associated with very slender and slender build are tuberculosis, pneumonia, "nervousness" and melancholia. The diseases associated with very fleshy or fleshy build are diabetes, nephritis and dropsy, apoplexy and arteriosclerosis and paralysis accompanying it; also numerous diseases of the alimentary tract. Incidentally, it appears that fleshy parents have, on the average, in their data, larger families than slender parents. Genetically, build seems to be controlled by multiple factors, with fleshiness tending slightly to dominate over slenderness. There is a marked tendency for persons of similar build (or with potentialities for such) to intermarry. Dissimilar builds are selected against.

THE CEREBELLUM OF THE FROG. O. LARSELL, J. Comp. Neurol. **36**:89, 1924.

The cerebellum of the frog develops from the rhomboidal lip of the fourth ventricle. The two lips grow laterally to form the auricular lobe and medially to become the body of the cerebellum. A swelling, the ventral cerebellar eminence, appears in the floor of the rostral portion of the lateral recess. This later fuses with the body of the cerebellum. The ventral eminence contains the nucleus cerebelli which sends fibers rostrally to form the brachium conjunctivum. This nucleus probably represents all of the deep cerebellar nuclei of higher forms. The cortex of the cerebellum possesses three layers; namely, a molecular layer, a Purkinje cell layer and a granular layer. Many of the cellular elements characteristic of the mammalian cortex cerebelli are present, but in a less specialized form. The most prominent bundle connected with the cerebellum is the tractus spinocerebellaris which enters the body of this organ; it corresponds to the tractus spinocerebellaris ventralis of mammals. Ascending branches of fibers of the eighth nerve pass to the cerebellum directly; also fibers from the vestibular nucleus enter the cerebellum. Both the direct and indirect vestibular tracts enter the auricular lobe. The fourth nerve sends some uncrossed fibers directly into the cerebellum. These fibers appear to be related to the mesencephalic fifth tract, which also passes through the cerebellum, and probably carry impulses of muscle sensibility. There is a direct tract, distinct from the fifth mesencephalic, from the roots of the fifth nerve to the cerebellum. Fibers also enter the cerebellum from the tectum and various nuclei within the medulla. In addition to the brachium conjunctivum, efferent fibers leave the ventral cerebellar eminence and other parts of the cerebellum for the tegmentum. The lateral line system, which disappears at metamorphosis, appears to be compensated for by the greater development of the vestibular system.

GRAY, Chicago.

A DESCRIPTION OF MATERIAL FROM A GYNANDROMORPH FOWL. MADGE THURLOW MACKLIN, J. Exper. Zool. **38**:355-376 (Nov. 20) 1923.

The body skeleton, head, feet, wing tips and the gonads of a gynandromorph fowl are described in detail. In life the bird appeared to be a hen, with neck feathering suggestive of the male and tail feathers longer than those of the

normal hen. The comb and right wattle were typical of the cock, and it exhibited male sexual behavior. It attempted copulation with apparent success. It was never heard crowing, and it did not fight other males. It was suspected of laying small eggs.

Every bone on the right side of the skeleton was larger than the corresponding bone on the left. The result was profound distortion of the axial skeleton. The spur, scales and toes of the right leg were larger than those of the left. The right cerebral hemisphere was larger than the left; the optic lobes seemed to be about equal; the right side of the medulla seemed larger than the left. The single testis appeared to be normal. Six ova were present, broken free from the ovary. A section of the testis showed well formed tubules with the lumen crowded with spermatozoa. The ovary consisted of a mass of ovarian tissue, through which was mixed more or less abnormal testicular tissue. Encapsulated in the center of the ovary was a large mass of suprarenal tissue. Masses of luteal cells were present in the theca interna and in the corpora lutea.

A lengthy discussion of similar cases and various theories follows the description. The author concludes that the fowl was a zygotic as well as a hormonic intersexual. The female hormone, supposed to function first, influenced the development of the individual toward the female side; the male, coming into play later, altered it in the direction of the male. The effects of both hormones, somewhat modified, are evident. The secretions of any endocrine glands were equally available to the two sides, so the explanation must rest on the different genetic constitution of the two halves, and because of this difference they each responded to their appropriate secretion and were less affected by the secretion of the opposite gonad. Apparently the soma on which these hormones work, plays a large rôle in the results. The bilateral asymmetry rests on the zygotic constitution of the individual.

WYMAN, Boston.

BRAIN ABSCESS, CLINICAL AND OPERATIVE DATA. CHARLES BAGLEY, JR., J. A. M. A. **81**:2161 (Dec. 29) 1923.

Brain abscess, still one of the most serious of cerebral lesions, requires prompt diagnosis and precision in treatment. The diagnosis often depends on some obscure sign or symptom. In answer to the usual question after all operations for brain abscess, "Do you believe the diagnosis could have been made earlier?" the author states that in the light of operative findings, one is always forced to acknowledge that more attention should have been paid to certain symptoms observed during the few days or even weeks before admission to the hospital.

The wide difference of opinion as to method in evacuating brain abscesses indicates the chaotic state of surgical procedure directed toward the relief of the condition. There is no one method applicable to all types of cases, but it is not likely that the chosen mode in a given type should vary from a small trephine opening to a large bone flap plus ventricular and lumbar punctures. After a study of the pathologic specimens and a full appreciation of the effort of the brain to surround the invading infection by a limiting membrane, Bagley developed a great respect for the abscess wall and determined to avoid any interference with its function. Operative procedures, therefore, must prevent any breaking of the abscess wall. In the usual type of deep brain abscess, the most simple drainage of the cavity through a small opening as near the bottom as possible gives the best results.

NIXON, San Francisco.

STUDIES ON THE RETINA. AN EXPERIMENTAL STUDY OF THE GECKO RETINA. S. R. DETWILER, J. Comp. Neurol. **36**:125, 1924.

The retina of *Gecko swinhonis,* a nocturnal lizard, was studied under conditions of light and dark adaptation. The retina of this animal consists exclusively of rods, which are arranged in regularly alternating rows of small, single and large, double forms. The inner segments of the rods contain a distal ellipsoid, a refractive paraboloid and a stout myoid. A refractive disk separates the inner and outer segments. The outer segment contains a granular, striated material which stains deeply with iron hematoxylin. The epithelial pigment projects down between the rods almost as far as their inner segments. During exposure to diffuse daylight this pigment was found to descend 5.6 microns. The normal retina is pink. Bleached isolated pieces of the retina do not regenerate the pink color, but if they are covered with the epithelial pigment, they will recover their colored substance on reexposure to darkness. Bleached isolated pieces of the retina left in situ will regenerate the color when placed in the dark again. Droplets resembling the striated material of the outer segments of the rods can be seen between the rods in abundance in eyes adapted to the dark. In such eyes the striated material with the rods is sparse. The converse situation exists in eyes adapted to light. Since their behavior corresponds with the regeneration and bleaching of the visual purple, some relationship between these histochemical substances and the visual purple is suspected.

GRAY, Chicago.

IMPORTANCE OF COMPLETE EXAMINATION OF THE CEREBROSPINAL FLUID IN SURGERY OF THE SPINAL CORD. WILLIAM J. MIXTER, J. A. M. A. **81**:2166 (Dec. 29) 1923.

The most important single sign of cord compression and the most reliable is that evidence which is obtained from the cerebrospinal fluid. At the Massachusetts General Hospital every patient suspected of cord tumor is prepared for puncture of the cisterna magna, as well as for spinal puncture, and lumbar puncture is performed. Pressure readings are made, pulse and respiratory oscillations are noted, and the patient is directed to cough and to draw a deep breath. Next, the jugular veins are compressed, and finally a small amount (usually 5 c.c.) of fluid is withdrawn. Notes are taken on the behavior of the fluid in the manometer during all these procedures. A portion of the fluid withdrawn is tested with alcohol for excess of protein. If the information obtained in these various ways gives definite evidence of block, puncture of the cisterna magna is not performed. If there is absolutely no evidence of block, puncture of the cisterna magna is not performed. If, however, there is the suspicion of block, either chemical or dynamic, the cisterna magna is punctured, combined readings are made, and the fluid is drawn from both needles for quantitative examinations of protein and for Wassermann examination and study of the cells.

NIXON, San Francisco.

THE SUBSTANTIA NIGRA AND ITS CONNECTIONS. C. FOIX and I. NICOLESCO, L'Encephale **18**:553 (Nov.) 1923.

This article is a histologic study of the substantia nigra, with special attention to its efferent pathways.

The authors comment on the value of comparative anatomy in enabling one to conceive the nature and function of the substantia nigra. Opinions have

varied as to whether it belongs to the mesencephalon or the diencephalon. Its morphology and its involvement in Parkinson's syndrome brings up the question of its relation to the globus pallidus. While the substantia nigra probably is related to both the cerebral cortex (Déjerine) and to the corpus striatum, the fact that in various mammals the pyramidal tract varies greatly in size while the substantia nigra has relatively the same volume, suggests that its extracortical relations are of primary importance.

The substantia nigra has two chief efferent pathways, one passing through the stratum intermedium, running toward the deeper aspect of the pyramidal tract and tending downward toward the bulb; and one whose fibers in part enter the posterior commissure. The first tract is probably in connection with lower centers controlling motor integrations, and its fibers may decussate. The latter tract is more difficult to understand. In the authors' opinion, it gives connection with the contralateral substantia nigra. By passing through the posterior commissure, it enters into the general commissural fibers—as is evidenced by the consistency and considerable development of this tract in various animals.

HYSLOP, New York.

LOCALIZATION OF BRAIN TUMORS BY DETERMINATION OF THE ELECTRICAL RESISTANCE OF THE GROWTH. FRANCIS C. GRANT, J. A. M. A. **81**:2169 (Dec. 29) 1923.

The author has tested the electrical resistance of normal dog and human brains, both fresh and hardened in formaldehyd; he also determined the resistance of pathologic brains. In a series of thirty formaldehyd hardened brains containing proved gliomatous tumors, the gliomatous tissue consistently produced a resistance of from one-half to two-thirds less than that of the adjacent normal tissue. In fifteen brains containing certified endotheliomas, the resistance of the tumor was a half less than the normal in twelve, in two from a third to a quarter less, and in one the resistance was approximately the same as the surrounding tissue.

At the operating table the author used the apparatus in twelve cases. Of these, five were gliomas. In every case, the resistance of the tumor was only half that of the normal brain. One was a sarcoma, and in this, the resistance between normal brain and tumor coincided closely with that of the gliomas. Of the endotheliomas, the resistance was lower than the normal brain tissue in four and higher in two.

NIXON, San Francisco.

Society Transactions

PHILADELPHIA NEUROLOGICAL SOCIETY

Regular Stated Meeting, March 25, 1924

CHARLES W. BURR, M.D., *President, in the Chair*

AN UNUSUAL CASE OF AMYOTROPHIC LATERAL SCLEROSIS. DR. M. A. BURNS.

The case is presented because of early development of symptoms with gradual return of function, although symptoms of anterior horn and lateral tract lesions are still present.

A youth, aged 19, a messenger, whose father and mother had died from tuberculosis, one sister having died in infancy, and who had one brother living and well, was a healthy boy until the age of 6, when, he says, he was struck by a heavy plank on the right shoulder. When between 6 and 8 years old, he was placed in a home for orphans and was told that his right arm and leg were not normal, and that he was unable to use his right hand to carry food to his mouth. After he had been in the home a short time a brace was applied to his right leg, which he wore for two years. He left the home when he was 13, and was able to walk fairly well without a brace, although he had little use of the right arm and hand.

Two years later, it was thought that the boy had tuberculosis, and he was sent to Mount Alto, remaining there for six months. There he received massage, but since that time he has received no treatment.

His chief complaint was partial paralysis of the right upper extremity and, to a less degree, of the right lower extremity, with marked muscular atrophy. He believed that in the last four years the arm and leg had improved.

Examination.—Station was good and gait was spastic on the right side. There was marked atrophy in the upper limbs, chiefly in the shoulder girdles, more marked on the right side. He had a tendency to a wrist drop on the right side and some vasomotor disturbance. No fibrillation could be demonstrated. Biceps and triceps reflexes were exaggerated on the right and slightly increased on the left. The grip on the right side was greatly diminished. Resistance to passive motion on the right side was poor, although it was fairly well preserved on the left.

There was marked atrophy of thigh and leg muscles on the right, especially in the gluteal region. The patellar reflex on the right was markedly increased and slightly overactive on the left. The Achilles reflex on the right was markedly increased; the Babinski sign was definite on the right, and very slight on the left. Ankle and patellar clonus were not obtained. All forms of sensation and the sphincters were normal.

Roentgen-Ray Findings.—One plate showed no evidence of an enlarged thymus. A second plate showed considerable cloudiness of the upper lobes of the lungs on both sides with a rather unusual type of peribronchial thickening. It is uncertain whether or not this was due to definite pulmonary pathology or to lack of inspiration.

Electrical Reactions.—The flexor muscles of the right upper extremity showed a normal response. The extensor muscles and muscles of the thumb showed a reaction of degeneration.

The serologic findings were negative, and the eye examination revealed nothing abnormal.

A Case of Unilateral Ascending Degeneration of the Pyramidal Tract. Dr. Francis X. Dercum.

A white, unmarried man, aged 39, was admitted to Jefferson Hospital, March 4, 1924, complaining of loss of power and stiffness in the right leg and right arm. The family history was unimportant. He had had gonorrhea about fifteen years before, and about the same time there was a local sore which healed promptly and which was not followed by any secondary manifestations.

The present condition began seventeen years before with slight weakness and stiffness in the right leg which gradually increased; later a sense of stiffness made its appearance in the right arm. This gradually became more pronounced. There was at no time pain or numbness.

Examination revealed a healthy-looking man with a gait suggestive of ordinary hemiplegia except that the spasticity in neither arm nor leg was as marked as that usually seen in typical hemiplegia following apoplexy. However, the arm was held automatically in a more or less semiflexed position, while the leg was held extended, and this was particularly noticeable in the gait. The face was not involved. The tendon reflexes in both the arm and leg were exaggerated. There was no Babinski sign, no wasting of muscles of the thenar or hypothenar eminence nor of the muscles of the shoulder girdle. There were no sensory losses. The right pupil measured 4.5 mm. and the left 5 mm. Both reacted promptly to light and convergence. The eyegrounds were normal. Serologically, blood and spinal fluid were normal.

This is the first case of the kind that I have ever observed. It is interesting to note that the onset of the condition antedated the local venereal lesion.

DISCUSSION

Dr. N. W. Winkelman: In Dr. Burns' patient, are the bones smaller on that side? It looks as if the hand is smaller, aside from the flesh. Could not the case be a developmental condition such as Little's disease?

Dr. Charles K. Mills: I believe that this case (Dr. Dercum's) of unilateral degeneration of the pyramidal tract is a real type of disease, one which I described some years ago. I saw one or two cases and another with Dr. Spiller, which came to necropsy and in which microscopic examination by Dr. Spiller showed degeneration of the pyramidal tract.

This type of disease must be carefully diagnosticated both from unilateral ascending paralysis agitans and amyotrophic lateral sclerosis of the ascending type. In one of my papers, I discussed at length the form of ascending and descending paralysis agitans.

Dr. Francis X. Dercum: I have never seen a case exactly like that of Dr. Burns'. There can be no doubt as to the structures involved, namely, the anterior cornua and the lateral tracts. Perhaps in the age and mode of onset lies the ultimate explanation.

I am satisfied that the case which I have just shown should be classified as one of unilateral ascending degeneration of the pyramidal tract; in other words, it is illustrative of the condition originally described by Dr. Mills.

Dr. M. A. Burns: I believe my case began as acute poliomyelitis in which the lateral tracts were evidently involved very early. The patient may have some shortening of his bone, but it is not marked. Most cases of Little's disease do not show atrophy of anterior horn cells, which apparently this boy has.

Cases of Endocrine Anomalies. Dr. Francis X. Dercum.

Case 1.—A white girl, aged 9, was admitted to the Jefferson Hospital because of mild Sydenham's chorea affecting mainly the left side.

The father had died at the age of 30 of meningitis. The mother was well and said that she had matured early and was adult in development at the time of puberty, which was established at 13. There was no history of mental, cardiorenal, pulmonary or malignant disease in the family.

The patient had had chickenpox and measles but no other childhood infections. She had been slow in learning to walk and talked plainly first at the age of 7. Very large tonsils and adenoids were removed at the age of 6. She was sent to school at that age, and made good progress. The mother said that the thyroid gland had for a long time been large, and that during the past year it had notably increased in size. The child had always been nervous and easily frightened.

At the age of 7, she began to menstruate, the periods recurring every twenty-eight days and lasting three, four and even seven days.

About a year before presentation, she was vaccinated, and two days later developed chorea, which improved greatly on rest in bed but did not entirely disappear.

The patient looked like a girl of 17. The thyroid gland was greatly enlarged and uniform. There was a marked mitral murmur; otherwise the visceral examination was of little importance. Gait, station and reflexes were normal. The mammary glands were well developed. The pubis and genitalia were adult in appearance.

There was no tachycardia or prominence of the eyeballs, and no von Graefe, Stelwag or Moebius sign. Pupils and eyegrounds were normal. The urine was normal. The blood contained 5,000,000 red cells and 7,400 white cells and 44 per cent. small mononuclears.

The child lay in bed playing with her doll and was easily pleased. Her talk and interests were childlike. Clearly there was no mental precocity. The basal metabolism was plus 9.

A roentgenogram of the skull revealed the posterior clinoid processes slightly larger than usual; otherwise the sella turcica appeared to be normal. The pineal gland was calcified. There was no roentgen-ray evidence of an enlarged thymus. But it must be borne in mind that the roentgen-ray examination of the thymus is very unsatisfactory, and that unless the enlargement is such as to cause marked shadows projecting beyond the sternum it cannot be observed. In this case, the large percentage of small mononuclears must be taken into account. It is to be regarded as evidence of lymphatic persistence, which naturally suggests also some degree of thymic persistence, and in the present instance would be in keeping with the history of tonsillar enlargement and adenoids; in other words, with an undoubted degree of lymphatic hyperplasia. If this interpretation of the case be correct, the precocious sexual development must be regarded as an expression of biologic failure. The enlargement of the thyroid must be regarded as an effort at

compensation. It is not accompanied by the symptoms of hyperthyroidism. Clearly it must be looked on as an *under*-compensation, and to prove the correctness of this view, I should state that thyroid administration has thus far been followed by a notable diminution in the size of the gland. This result is further in keeping with previous observations of my own that thyroid enlargement in girls at or near puberty is the expression of an effort at compensation. Commonly such enlargements disappear on thyroid administration. If hyperthyroidism, the opposite condition, were present, such a result would assuredly not ensue.

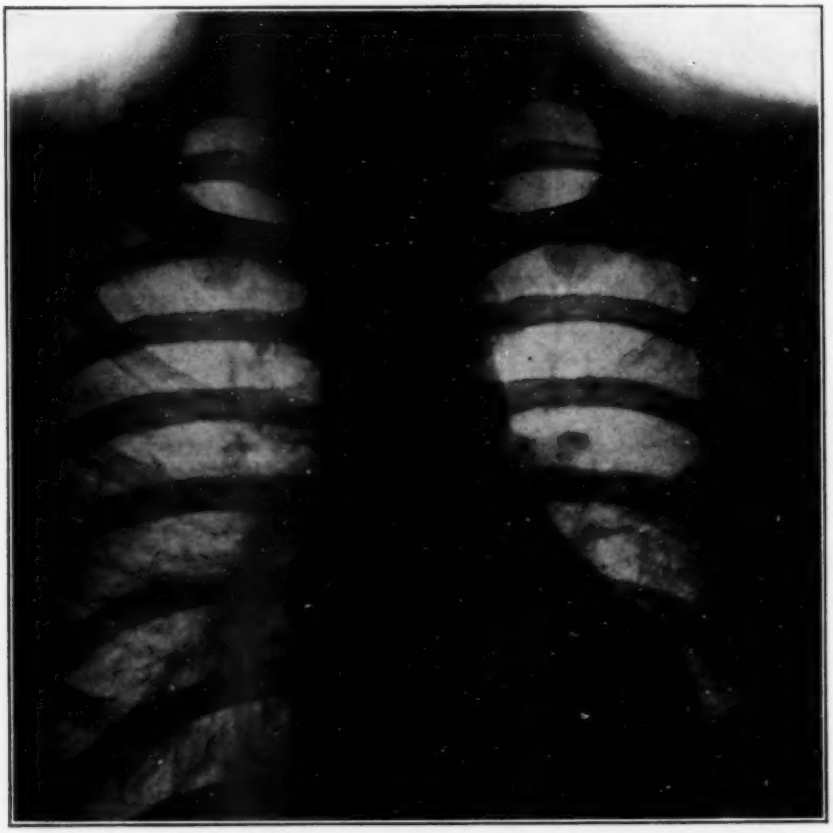

Fig. 1 (Case 2).—Enlargement of the thymus, especially marked in the supraclavicular region.

Case 2.—A Hungarian woman, aged 21, single, a cigar-maker, was admitted to the Jefferson Hospital, March 21, 1924. She presented herself first at the outpatient department suffering from an ordinary Bell's palsy on the right side. It was noted also that she had decided enlargement of the thyroid gland. The family history was negative except that one sister presented thyroid enlargement similar to that of the patient. The personal history was negative as regards childhood infections, with the exception of measles. Menstruation

appeared at 16, was painful and persisted for a year, after which it ceased for seven months. Since then it had been normal. The facial palsy had been present for one week. She said that she had not been well for four years.

She was rather thin and awkward, 5 feet 7 inches in height. The viscera were normal. The marked thyroid enlargement was symmetrical and soft. There was no bruit over it. No adenopathy was detected. Gait, station and reflexes were normal. The Wassermann tests of the blood and spinal fluid were negative. The urine showed a faint trace of albumin; otherwise it was normal. The differential blood count showed 27 per cent. small mononuclears. The eyegrounds, pupils and ocular muscles were normal, except that there was weakness of the right orbicularis incident to the Bell's palsy.

Fig. 2 (Case 2).—Large pituitary fossa.

The patient said that the thyroid enlargement was at one time much more marked, that it became decidedly less during her voyage to this country, and that she suffered from enlarged lymphatic glands but that these improved under treatment.

The roentgen-ray examination revealed enlarged thymus extending not only into the costal interspaces, but also into the supraclavicular area and a pituitary fossa somewhat larger than the average (Figs. 1 and 2).

The enlargement of the thyroid gland I think must be regarded as compensatory. It is probable that there was also enlargement, moderate in degree, of the anterior lobe of the pituitary. This was suggested by the patient's height, and, if so, like the enlargement of the thyroid is to be interpreted as an effort at compensation. Both are to be regarded as under-compensations. There was no tachycardia, no exophthalmos, no von Graefe or other ocular

phenomena. The basal metabolism was recorded as plus 1. Here again I believe thyroid administration is indicated.

CASE 3.—A white boy, aged 9, had an unimportant family history, except that a sister, aged 3, had a rightsided hemiplegia. The patient, who was the first born, was a full term child. Birth had been normal. He was breast fed. He learned to talk when a little over a year; he walked at 2 years of age. He frequently had vesical incontinence, and still occasionally soiled his clothing. About six weeks before presentation, the adenoids and tonsils were removed.

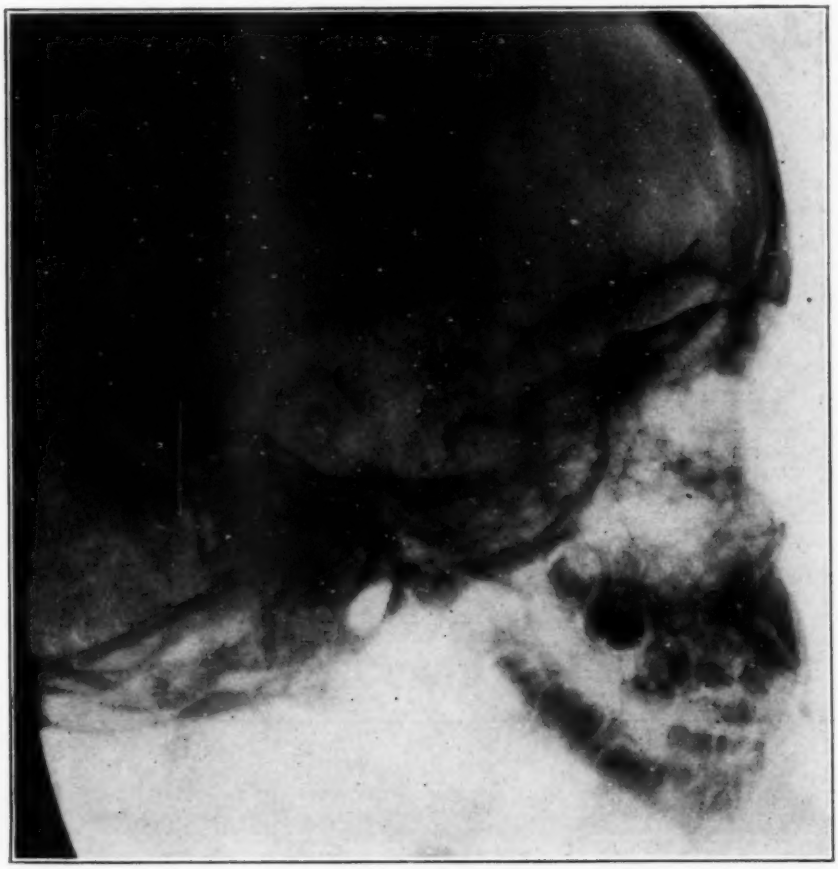

Fig. 3 (Case 3).—A much enlarged sella turcica.

At the fourth year the mother noticed that he was not growing, and said that he had been growing slowly since.

He was 38 inches in height. The trunk, arms and limbs appeared to be properly proportioned. The head, general skeletal and muscular development revealed no peculiarities. The ears, hard palate and teeth were normal. Station, gait and reflexes were normal. The pupils were normal. Speech was infantile and rather slow. He was very quiet, took little part in the play of other children and was mentally much retarded.

The viscera and thyroid were normal. There was no infiltration of the skin and no evidence of cretinism. Roentgen-ray examination of the chest revealed a widening of the cardiac shadow at the base of the heart on both sides, evidently due to a persistent thymus. Roentgen-ray examination of the head revealed a sella definitely larger than the average. It was almost twice the size for a child of his age (Figs. 3 and 4).

The case appears to be one of dwarfism featured by a persistent infantilism. The significance of the enlarged and persistent thymus is unmistakable. For some reason there was no compensatory enlargement of the thyroid. The enlargement of the pituitary, however, I regard as of great significance. I

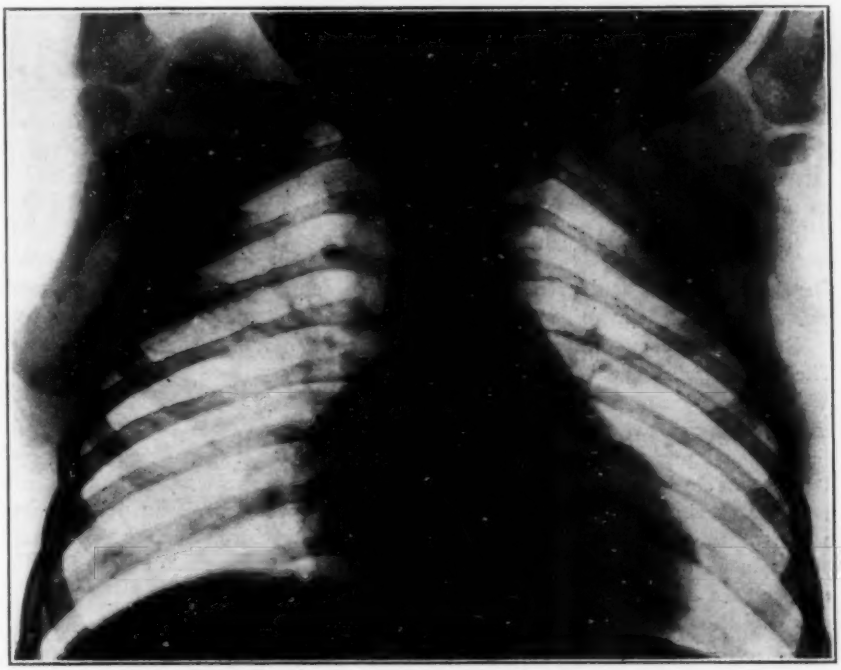

Fig. 4 (Case 3).—Decided enlargement of the thymus in a child aged 9.

believe that the anterior lobe of the pituitary stands in a similar relation to the thymus as does the thyroid. Thymus persistence is commonly associated with thyroid increase, and sometimes appears with anterior lobe increase alone, as in the present case. The question at once arises. Why did not increased stature instead of dwarfism result? Evidently we are dealing with a failure of compensation; the pituitary enlargement is an *under*-compensation; it is vastly overweighted, so to speak, by the enlarged and persistent thymus. The latter again is in keeping with the tonsillar and adenoid history of the patient. As before, the clinical picture is that of a biologic failure.

Case 4.—A sister of the previous patient, aged 3, also presented endocrine features. Her birth was normal. She was bottle fed, but was well until 14 months of age, when her right arm was scalded, for which she was in a hospital

for seven weeks. When she left the hospital, it was found that the right arm and leg were weak. It was four months before she walked, when it was noticed that she dragged her right leg.

There were scars of a burn over the inner aspect of the upper arm and right side of the chest. The right arm and leg were slightly spastic; the right knee-jerk was plus compared with the left, and a Babinski sign was present on the right. A roentgenogram of the chest revealed some enlargement of the thymus. A roentgenogram of the head was negative. The case was interesting because of the thymus enlargement. That other glandular imbalances will develop later is improbable. The cause of the hemiplegia is unknown. An embolism of course suggests itself, although the heart was normal. The enlargement of the thymus is interesting because of the same condition in the brother.

Case 5.—A girl, aged 2, whose birth was delayed and instrumental, who did not take the breast, cried very little and seemed heavy and stupid for about two weeks, when one month old suffered from a Bell's palsy, cried a good deal, was difficult to feed and did not gain in weight. When 3 months of age, she developed pyelitis. She did not take notice of anything until she was 8 months of age, when she began to notice moving objects, but she never attempted to grasp or reach at them. A roentgenogram of the head taken at 14 months revealed a moderate degree of hydrocephalus. She cries little now and is still difficult to feed.

The palate was high but the lobes of the ears and pinnae were well formed. Dentition was much delayed. The pupils were normal. The anterior fontanel was open. Wassermann examination of the blood and spinal fluid was negative.

Roentgen-ray examination of the chest revealed a much enlarged thymus. The sella turcica and thyroid gland were normal.

The history suggests intracranial injury at birth. The absence, however, of convulsive seizures and of hemiplegic or diplegic symptoms shows that gross hemorrhages probably did not occur. Of course the roentgenogram suggesting a moderate hydrocephalus at 14 months must be borne in mind, and yet one cannot help ascribing a special significance to the very large thymus. Sajous has many times emphasized the relation between retardation or arrest of mental development with enlargement of the thymus, and the present case is in keeping with such a relationship.

Case 6.—A girl, aged 11 months, an only child whose parents were well, was a full term baby and breast fed. At four months the mother noticed that the eyes were being rotated from side to side. Both legs were somewhat spastic, with a bilateral Babinski sign. The palmar reflex was still present and vigorous. The anterior fontanel was normal. The pupils were normal. Rotation of globes was not a true nystagmus and was only present at times. She paid no attention to bright or moving objects and seemed dull, heavy and indifferent.

A roentgenogram of the chest revealed an enlargement of considerable size in the right upper mediastinum, which was apparently an enlarged thymus. A roentgenogram of the skull revealed the bones to be rather thin, and a normal sella.

Unfortunately, we do not possess satisfactory means for studying the thymus during life. In children more satisfactory roentgenograms of the thymus can as a rule be obtained than in adults. In the latter, the thymus must be decidedly enlarged before shadows appear beyond the sternal edge in the costal interspaces.

That the blood reveals, at least at times, an excessive percentage of lymphocytes is most suggestive and interesting. Many years ago I frequently noted in making blood counts in cases of neurasthenia also an excess in the lymphocyte count. Of course I noted from time to time morphologic peculiarities in neurasthenics; moderate evidences of arrest or deviation, sometimes peculiarities of the circulatory apparatus, such as heart and vessels that seemed rather small and perhaps poorly developed, but the excess of the lymphocyte count I could not understand. Is it possible that its significance is that of a slight or moderate thymic and lymphatic persistence, a condition which expresses in slight or moderate degree that which we find typically expressed in a status thymico-lymphaticus? Perhaps we have here an explanation of the feebleness of resistance to fatigue in neurasthenic cases.

DISCUSSION

Dr. B. P. Weiss: I think Dr. Dercum's interpretation of the association of the thymus with the thyroid is valuable. In the huge number of tonsillectomies performed, occasionally an unexplainable death occurs which to my mind may be due to an enlarged thymus. The nose and throat men should be informed of the frequency of a persistent thymus in early life.

Dr. Francis X. Dercum: I have elsewhere emphasized the relationship which exists between the thymus, the thyroid and the pituitary glands. Both the development of the thyroid and the development of the anterior lobe of the pituitary appear to be related to the development of the thymus. The series of cases I have presented is therefore of great interest.

A Case of Chronic Progressive Ophthalmoplegia of Forty-Five Years' Duration (Wilbrand and Saenger Type). Dr. W. B. Cadwalader and Dr. H. Maxwell Langdon.

A widow, aged 81, had had drooping of the left lid for forty-two years; the right lid began to droop shortly after the left. There was now complete bilateral ptosis with inability to move the eyes, except the left slightly upward. The pupils were equal and responded normally to light. The lenses were slightly hazy, the arteries moderately sclerosed; vision of the right eye was 5/20 and of the left 5/6; the visual fields were normal. Neurologic examination revealed no changes except moderate arteriosclerosis and slight atrophy of the left half of the tongue.

The case was one either of degeneration of the nuclei of the third, fourth and sixth nerves or of a form of abiotrophy (Gowers) which may be either primary nuclear degeneration or primary muscular degeneration. Microscopic examination of the muscles in a somewhat similar case made by Fuchs, in 1890, showed the fibers atrophic with pigmentary degeneration inside the sarcolemma. Hence he thought it was a primary muscular atrophy. Wilbrand and Saenger have reports of thirty-two similar cases; six cases have been reported since then, making thirty-eight, and this case is the thirty-ninth on record so far. There has been no microscopic examination of the nuclei in such a case. Were the nuclei the site of the trouble, it seems probable that the pupillary portion of the third nerve nucleus would at some time be involved, but all of these patients have had normal pupillary reaction to light. There is quite a distance between the third and sixth nuclei. If the condition is nuclear, it seems strange that some intervening parts have not been involved.

DISCUSSION

DR. N. W. WINKELMAN: Anatomists state that the third, fourth, sixth and twelfth nuclei have the same relation to the central canal as the anterior horn cells, and they represent the continuation of the large anterior horn cells. They are derived from the myotomes. I think it is therefore not a question of proximity but of development and relation.

A CASE OF CAUDA EQUINA LESION DUE TO INTRASPINAL TREATMENT. DR. C. A. PATTEN.

A man, aged 37, single, gave a negative history, except that he had had a Neisserian infection on three occasions, and ten years ago had a primary syphilitic lesion for which he received no treatment except the local use of a dusting powder. There were no secondary manifestations, and the patient was well until July, 1923, when he noticed that his ankles were swollen, that he was short of breath, and that his heart "beat like a trip hammer" on exertion. He was in bed three weeks, and practically recovered.

In November, 1923, he experienced precordial pain, and thought he would collapse. His physician told him that he had an enlarged heart, and referred him to another physician who told him that he had neurosyphilis and advised treatment. Before treatment, the blood and spinal fluid were examined and found negative. Another examination revealed a negative blood but a three plus Wassermann reaction of the spinal fluid. There were no other findings. He then received three or four intravenous injections of neo-arsphenamin, which extended into December, 1923. At the end of this time, all serologic tests were reported negative. At the time of the fifth intravenous injection (December, 1923) the patient said that "The doctor put the same yellowish-green stuff into my back." This intraspinal treatment was given while he was sitting in a chair, and was the first of its kind. As near as can be learned, an autoarsphenamized serum was not used, but probably a diluted neo-arsphenamin solution, a part of the same solution that had been injected into his arm. Immediately following the intraspinal injection, the patient fell to the floor in collapse because of the severe pain in the back. He lay on a table for an hour, and then walked around for several hours, the pain in his back being intense. He described the pain as an intense burning sensation which did not radiate. His legs felt numb and weak. Gradually the legs became weaker, until on the third day he could not stand. For two weeks, there was total paralysis of the lower extremities, at the end of which time he could move his legs a little, and after exercising in bed for about two weeks he was able to walk a little, but he reeled like a drunken man. His legs gradually became weak again, and since that time he had not been able to walk or stand. There had been incontinence of urine and feces since the date of the intraspinal injection, and the numb feeling in the lower extremities had persisted without change.

In January, 1924, he was sent to a hospital where a spinal fluid examination showed 80 cells, mostly lymphocytes, and a colloidal gold curve suggestive of syphilis; otherwise it was negative. Cystitis was present at this time, probably due to retention.

Since admission to the Orthopaedic Hospital in the service of Dr. Weisenburg early in February, the patient had been unable to walk or stand, but had been able to move his legs in bed. Incontinence of both urine and feces had persisted, and the abdomen had been constantly distended. The muscles of the

lower extremities were wasted, the knee jerks were prompt, and the ankle jerks absent. There was no Babinski sign. There were occasionally twitchings of the thigh and leg muscles, but no fibrillation, and sometimes when the plantar surfaces were stimulated, the lower extremities jerked up in a sort of spasm.

There was complete loss of sensation in the fourth and fifth sacral segments, gradually fading out into the third, second and first segments, a saddle-shaped area around the anus, buttocks, perineum, genitalia and slightly on the postero-internal surface of the thigh. Pain and tactile sensibilty were considerably impaired up to the level of the great trochanters, bilaterally, and gradually shaded into normal at about the level of the sixth dorsal segment. Vibration sense was markedly impaired but not lost. Sense of position was lost in both feet. There was marked ataxia of both lower extremities. Electrical examinations revealed no reaction of degeneration, although the responses were not quite normal in quantity. The upper extremities were normal.

The spinal fluid showed three cells and a two plus globulin reaction; otherwise it was negative. The urine contained albumin and pus. The blood showed 12,400 leukocytes. The blood chemistry was normal in all respects.

Comment.—1. The case history and serologic reports do not substantiate a diagnosis of neurosyphilis; the clinical symptoms since July, 1923, have pointed to a visceral rather than to a nervous system lesion.

2. Even granting the possibility of neurosyphilis, intraspinal treatment was not indicated, at least until the usual safer methods had been given a fair trial. I am fully convinced that intraspinal therapy with arsenic and mercury should be employed only when all other methods have failed. A physician friend has related the following: Five young men, all actively and efficiently engaged in business were found on spinal fluid examination to have neurosyphilis. They were hospitalized immediately, and on the same day each received an intraspinous treatment of arsphenamin in horse serum. One died in forty-eight hours, and one a month later. Of the three who survived, one had spastic and two ataxic paraplegia, the latter having total incontinence. In the following two years one of the three died, and one committed suicide. The third is still living, able to walk with the aid of canes and to conduct his business in a small way. The moral is obvious. Too many enthusiastic persons believe that they are competent to treat the nervous system, and "fools rush in where angels fear to tread."

3. The symptoms and neurologic findings in this case indicate at present definite involvement of the spinal nerve roots, the brunt of the toxic irritation being borne by the sacral nerves. The patient received the treatment while sitting, and for some hours afterward maintained an upright posture, and either through the force of gravity or by reason of the lymph drainage along the nerve sheaths, those nerve roots that have the lowest exit from the canal were more intensely affected. There was undoubtedly some diffusion upward, as evidenced by the mild sensory disturbance up to the dorsal segmental area.

4. The outcome I believe is bad, because the nerves have been too severly damaged to recover. There is probably an extension beyond the spinal canal along the nerves.

A Case of Psychoneurosis. Dr. James J. Waygood.

A man, aged 44, was brought to the Pennsylvania hospital, because the previous night, Christmas night, he had left home "to keep an important engage-

ment," had not returned and had been found in the morning a short distance from his home lying on the ground covered with frost, and with his hat, coat and overcoat removed. On admission to the hospital, he refused nourishment, refused to speak and to allow his eyes to be opened, although he appeared to be aware of his surroundings.

Physical examination and laboratory findings were all negative. The family history was merely suggestive. One sister was said to have been "hysterical" for a period of two years, and the father was an irritable, domineering type of man. The patient was the first child and only son in a family of three, a delicate child, slow in developing and suffered from headaches and stomach trouble until he was a young man. His physical condition had improved following appendectomy.

He did well in school, but was shy and did not mix with other boys. His interests were unusual, and later he developed along the lines of mechanical and manual work. He grew up with a strong mother attachment, and felt that his father was opposed to him. In spite of this, following the death of his mother, when he was 22, he entered business with his father, but the relationship was not a happy one, and later, after his own marriage, he broke completely with his father. From this point on there was a strong motive in the patient's ambition to outdo his father in success.

In recent years, the patient had had several positions, had been fairly successful and frequently on the fringe of big business, but each time, in some part due to his own personality, conditions had become intolerable to him, and he had been forced to withdraw without any definite plans for the future. He was forced to resign from his last position a year before the present illness. He speculated in stocks and made some money, then lost some, stopped, and spent the latter part of the year in close study of economics and finance, of which he already had considerable knowledge, feeling that he could solve the laws which governed the securities markets.

The onset of his illness was abrupt. He was in good health. There was no element of exhaustion. For a few days there was complaint of dulness in the ears. Two days before Christmas he attended church with his wife, and following this appeared to be mildly excited; he had a long talk with the minister on religious topics. On Christmas eve, he was excitable while trimming the Christmas tree, and later in the evening left the house, saying that he was going to seek the Lord. He returned at 3:30 a. m., and retired quietly. On Christmas morning, he said that he had found peace. During the day his behavior was natural.

In the hospital, he lay for three days quietly and comfortably in bed. He required tube feeding, to which he was mildly resistant, and during the first feeding he prayed loudly and devoutly. His expression was alert and attentive, with an occasional smile. Unexpected stimuli caused him to start. On the second day, he occasionally answered a question with a nod or a shake of the head. On the third day, he began to talk a little, remembered the physicians' names, and said that his eyes would be opened at the right time, that God would show him his light and raise his eyelids. The following morning, he arose, ate an excellent breakfast and talked freely. His mood was moderately exhilarated. He was anxious to talk.

His experience as described by him was a religious one. He was under the absolute domination of the Lord. There were no long lapses of consciousness. He was "racked from head to foot" with cold, realized where he was,

what was being done for him, but was "in perfect composure" and "wished only to be left alone." For two days he talked freely of his business difficulties, did a good deal of preaching and advanced some peculiar ideas. He felt that for the past year a tremendous religious revival had been taking place in this city, equal to that of the Billy Sunday campaign in its scope, conducted by such men as McCartney and Norwood, and that in this campaign the radio and the Ku Klux Klan had been utilized. He believed that the security markets were controlled by a clique, who used various methods to determine their unified action. Their activities were guided by signs of the Zodiac. Various comic strips in the newspapers were used to broadcast their purposes.

At this point he insisted that he had fully recovered, and demanded his release. His agony was extreme, as he insisted that the physicians were taking pleasure in crushing his spirit and in watching his sufferings. When interviews were discontinued, he adjusted himself and took pleasure in the hospital activities, but when two weeks later another attempt was made to reach his difficulties, the same reaction occurred, and he left the hospital. He had a six weeks' enjoyable vacation on the Pacific coast, and on his return he was full of ambition to tackle the job of finding work. He was last seen a week later, and was most concerned and somewhat disheartened at his inability to find the kind of work he wanted. He wanted to be independent, not at the beck and call of some one else. He talked of speculative real estate building. He still retained the peculiar ideas regarding the religious revival and the control of the security markets. His self-sufficiency, insistence on his own opinion, and inability to accept advice were outstanding features during his convalescence, and are still most prominent.

Two Brains Showing the Lesions Producing Cerebral Monoplegia. Dr. N. W. Winkelman.

The first brain is from a man aged 70, admitted to the Philadelphia Hospital, Feb. 8, 1924. He died fifteen days later. The patient complained of paralysis of the right side of the face and difficulty in speaking. The onset of this condition was sudden about fifteen years ago, and since that time his speech had been more or less stuttering.

On admission he had marked weakness of the lower part of the right side of the face and right side of the palate. Speech was definitely stuttering, and not aphasic. He developed gangrene of the legs, from which he died.

The brain showed an old area of softening 2.5 cm. long and 1.5 cm. wide, sharply limited to the lowermost part of the left motor area, not involving the frontal convolutions not penetrating beyond the external capsule and not involving the frontal lobe.

The second brain was from a colored woman, aged 38, admitted to the Philadelphia Hospital for the first time on July 21, 1921, with a history of having suddenly developed paralysis with numbness and tingling of the left lower extremity. Six months later she had another "stroke," with weakness of the entire left side, which had remained, but the lower extremity had always been more involved than the upper.

The brain showed an area of softening limited sharply to the paracentral lobule; the thrombotic lesion of the anterior cerebral artery could easily be made out. There was another lesion in the internal capsule on the same side, which explained the hemiplegia.

CEREBRAL SEGMENTAL MONOPLEGIA. DR. A. M. ORNSTEEN.

Three patients with limited cortical disease resulting in segmental paralysis and anesthesia were shown. In two of the cases a cursory examination might lead the examiner to a diagnosis of peripheral neuron disease; in one, this error had actually occurred.

CASE 1.—A man had paralysis of the fingers of the right hand dating from a dislocation of the right shoulder five years before presentation. A brachial plexus injury might be indicated, but the history and examination soon dispelled this idea. In the accident referred to, he was struck on the left parietal region by a falling iron bar. He was not unconscious, but as he fell the right arm shot up above his head, made three rather slow revolutions and fell helpless to his side (a jacksonian manifestation). With assistance he was able to walk a short distance to receive first aid, which consisted of general anesthesia and reduction of a supposedly dislocated right shoulder with the application of a fixation dressing; no attention was given to a slight scalp wound. He then rode home in an automobile and was able to undress and go to bed without assistance, one and one-half hours after the accident. Apparently, he went to sleep, but for three days he could not be awakened. On regaining consciousness, he had complete paralysis of the right leg and motor aphasia; the arm was in a splint and probably was also paralyzed. In about three weeks, he was able to speak quite well and in six he was able to walk. At about this time, the splint was removed, and the arm was found to be completely paralyzed. During the period of complete paralysis of the arm, automatic associated movement occurred several times in the form of strong flexion of the paralyzed arm during yawning (pandiculation or stretching reflex). Gradual improvement of the arm took place, until now he can use the whole limb normally except the hand, which is spastic, and all movements of the hand and fingers are weak and awkward, especially in making a fist and in adduction and abduction of the fingers. There is loss of sense of position and passive motion in the fourth and fifth fingers; the recognition of compass points is greatly impaired in all the fingers and the hand. Astereognosis is not present, but the patient says that several years ago he had difficulty in recognizing objects in his right hand. There is no appreciable difference between the innervation of the two sides of the face, and the tongue does not deviate. The deep reflexes of the right side are exaggerated, but no extension of the great toe occurs. Sensation in the lower limb is intact. A small depression in the skull is present over the motor area for the upper limb on the left side. The interpretation given to this case is a segmental monoplegia of the right hand from local injury of the cortex (depression of the skull), and a middle meningeal hemorrhage producing the previous hemiplegia and motor aphasia.

CASE 2.—A man, aged 69, while at work eleven days before presentation became dizzy, nauseated and experienced numbness in the right foot, which felt as if it were off the floor. The weakness in the foot progressed, and the next day it was completely paralyzed. There was some weakness in the knee, but much less than in the foot. Sense of position and passive motion were not disturbed. There was a Babinski sign, and the knee jerk was exaggerated; the ankle jerk had almost disappeared. Here we have a segmental monoplegia affecting the right foot from a thrombotic lesion of the left anterior cerebral artery at its extreme and affecting the left paracentral lobule.

CASE 3.—A man who had been a painter for thirty years recently began to complain of numbness in the left hand and foot, which was ascribed to lead

neuritis. Investigation revealed the cortical nature of the symptoms. Five months before, while going up stairs, he suddenly experienced numbness in the fingers of the left hand; a few hours later numbness appeared in the lower half of the left side of the face, on the inner side of the forearm, arm and over the precordium; and in six hours the same symptom appeared in the toes of the left foot. This condition had persisted unchanged. There were no other general or local symptoms. The reflexes on the left side were all exaggerated with flexor toe response. There was slight hypesthesia and hypalgesia in the fingers and side of the face affected. Astereognosis in the left hand was complete, with defect in sense of position in the toes of the left foot. There was no weakness in the affected limbs, nor were there any sensory phenomena either subjective or objective in any other part of the affected side. The blood pressure was: systolic, 280; diastolic, 160; the radials and temporals and retinal vessels were sclerotic. Here again there had occurred a thrombosis of a cortical vessel (middle cerebral) producing segmental sensory changes; in the arm, radicular in distribution.

Obstetrical Brachial Paralysis with Involvement of the Cervical Sympathetic Nerves. Dr. W. B. Cadwalader.

A white child, 2 years old, was born in difficult labor, the details of which could not be obtained. The mother said, however, that the right arm was paralyzed at birth. Examination revealed complete paralysis of all muscles of the right upper extremity. Sensation apparently was lost throughout the hand and the forearm, not in the upper arm. Dr. Ornsteen found complete reaction of degeneration of the deltoid, biceps, triceps, radial extensors, finger extensors, and the ulnar and median group of muscles. The right pupil and the right palpebral fissure were smaller than the left. Roentgen-ray examination showed marked lack of development of the upper epiphysis of the right humerus.

I venture to call attention to the involvement of the cervical sympathetic nerves, indicating that the root emerging from the first thoracic segment must be involved. This could easily occur if there had been severe traction of the head at birth and the shoulder had been obstructed against the symphysis. In this way, all the nerve roots of the brachial plexus could be implicated.

Hydrocephalus Secondary to a Cerebellar Tumor Causing Cystlike Distention of the Third Ventricle, with Symptoms of Pressure in the Region of the Pituitary Body. Dr. John H. W. Rhein.

A boy, aged 18, who was admitted to the Polyclinic Hospital on Oct. 13, 1922, with a negative family history, had had a sunstroke when 5 years of age and influenza and pneumonia in 1918. He had always been mentally deficient. He had little eduction, had not advanced beyond the third grade, but otherwise had been apparently perfectly well and healthy. Two years previously, vision began to fail, and he was completely blind in February, 1922.

He was a large boy with a marked deposit of adipose tissue over the entire body. The genital organs were normal. The growth of hair was profuse, and the pubic hair was masculine in type. There was no paralysis in the facial distributions; the tongue protruded in the median line; the masseter muscles contracted equally well on both sides; the abdominal, cremasteric and epigastric reflexes were active. The arm reflexes were diminished; the knee reflexes were equal and increased, the Achilles normal on both sides, and persistent clonus

was present. Oppenheim and Babinski reflexes were absent. The sense of smell was diminished on the left side. The anteroposterior diameter of the head was greater than the lateral diameter. The Romberg sign was negative. With the feet together or even slightly separated, a push on the chest would make him fall backward. On two occasions after admission to the hospital, he had two or three slight general convulsive seizures. He was apparently conscious in these, and the attack was followed by headache.

The urine was negative; the blood count showed no abnormalities, and the Wassermann reaction was negative. The cerebrospinal fluid showed two cells per cubic millimeter, a negative Wassermann reaction, protein, globulin and sugar, and the colloidal gold curve was 1112221000.

The sugar tolerance test was diminished: 0, 87 mg. per 100 cubic centimeters; 1, 210 mg. per 100 cubic centimeters; 2, 142 mg. per 100 cubic centimeters; 3, 110 mg. per 100 cubic centimeters. The blood chemistry revealed no abnormalities, and the basal metabolism rate was minus 11.

The roentgen-ray examination by Dr. Pfahler showed apparently partial destruction of the anterior and posterior clinoid processes. It is difficult to outline the sella even with the stereoscope.

The sinuses were examined with the roentgen ray, but except that the frontal sinus on the left was not seen and on the right was quite small, no disease was found. Dr. Ralph Butler gave the following report: There was a slight deflection to the left of the septum and a thick discharge on both sides. The blood vessels were enlarged on the septum on the right side; the tonsils hypertrophied and submerged, and pus could be expressed from them.

Examination by Dr. Peters revealed large, equal, regular pupils, which did not respond to light. The right disk was pale, infiltrated near the edge, with very much blurred arteries. There were small and contracted macular twigs, almost ropelike in tortuosity, twisted like a knot in a rope, one third the size of the veins. There was no perivasculitis. On the left side, the edge of the disk was obliterated in spots, the disk white and chalky. The vessels were lost in this white exudate, and the arteries were very small, ropelike and tortuous, and were completely obliterated in spots, showing in other areas white lines. There were numerous atrophic areas in the retina. The diagnosis was atrophy secondary to neuroretinitis. There had possibly been choked disk some time on the left, and the primary condition was undoubtedly violently inflammatory.

Examination by Dr. Roberts revealed nerve deafness on the right. The Bárány test made by Dr. Fisher indicated some pressure on the fourth ventricle showing cerebellar involvement.

Dr. Grant operated, attempting a trans-sphenoidal approach. The patient died seven hours after an unsuccessful efferot to reach the pituitary region.

The brain contained a tumor in the right hemisphere of the cerebellum. There was pronounced hydrocephalus, especially on the left. The third ventricle which measured 2 cm. by 2 cm. by 2.5 cm., presented the appearance of a cyst at the base of the brain. This cyst had so compressed the optic chiasm as to reduce it to a very thin band. It was also sufficiently large to have compressed the hypophysis. The pituitary body was not enlarged, and the clinoid processes were necrosed.

The symptoms of note were moderate adiposity, lowered sugar tolerance, epileptiform disturbance and atrophy of the optic nerves, secondary in type. The cerebellar symptoms, if any, were overshadowed by those relating to the

pituitary body and sella. The only symptom which suggested cerebellar involvement was a tendency to fall backward.

This case belongs to a group of cases which have been reported, the symptoms arising from hydrocephalus, which causes cystic formations in various fields of the brain.

I have reported such a case in which cystic formations at the cerebellar pontile angle gave rise to cerebellar symptoms, and there are on record three cases reported by Schultz and Stumpf, in which hydrocephalus causing pressure on the hypophysis produced adiposity and hypoplasia of the generative organs. These and the ones reported by Cushing and Strauch are the only ones that I have found in the literature.

DISCUSSION

Dr. Francis Grant: At necropsy, the sella turcica was found to be definitely enlarged, and both anterior and posterior clinoid processes had been worn away. This case illustrates again that it is often difficult to distinguish between pituitary and cerebellar lesions. I believe that we have encountered at least six cases in which the clinoid processes have been eroded by internal hydrocephalus resulting from cerebellar disease, and in which the lesion was subtentorial and not supratentorial. The Bárány test is of great importance, for it may distinguish between a cerebellar and a pituitary lesion. When the roentgenogram shows erosion of the anterior and posterior clinoid processes, unless other definite evidence of pituitary disease exists, and there is no evidence of a large extrasellar growth, we always suspect that there are lesions in the posterior fossa.

CHICAGO NEUROLOGICAL SOCIETY

Annual Meeting, May 17, 1924

Julius Grinker, M.D., *President, in the Chair*

Epileptiform Catatonia. Dr. M. Urstein (Warsaw, Poland) by invitation.

A group of patients was presented who had been subject to epileptic seizures for many years. Marked improvement and cessation of fits had been observed under treatment with various glandular extracts during a period of about six weeks. All these patients showed certain catatonic features which were regarded by the author as indications that the fundamental disease was catatonia.

Motor Responses to Sensory Stimuli in Infants. Dr. Mandel Sherman.

The purpose of the study was to observe a number of sensorimotor responses in normal full term infants varying in age from 1 hour to 12 days. The particular reactions studied were the plantar and pupillary reflexes, the response to sticking with a needle, the early habit formation of defense movements of the arms and the coordination of the eye muscles. The study was made on ninety-six infants at the Chicago Lying-In Hospital. Uniform conditions were obtained during the examinations. The infants were stripped of clothing and laid on a hard cushioned flat table. The room was darkened when examining for the pupillary reflex and for coordination of the eyeballs. The light used for the pupillary reflex was just strong enough to produce a prompt response in the

adult eye. A weak light was used as a stimulus when coordination of the eye muscles was studied.

The results of the various reactions studied were:

1. *Response of the Pupil to Light.*—Pupillary reactions were classified as fixed, very sluggish, sluggish, and good, according to the rapidity and amount of contraction. The reaction of the pupil became increasingly adequate, both in amount and rapidity, with advance in age. The mean age of the subjects with fixed pupils was 1.8 hours; of those with very sluggish pupils, 7.7 hours; of the children with sluggish pupils, 19.8 hours. The reaction of the pupils in all cases above 33.5 hours was good. The results indicate that under controlled conditions the pupillary reaction is present in normal infants from about the age of 3 hours on, and becomes increasingly adequate up to 30 hours of age.

2. *The Plantar Reflex.*—This reflex was elicited by stroking with a dull pointed pencil from the anterior plantar surface. When the stimulus was followed by extension of the great toe, five additional stimuli were given in succession. Flexion of the toes occurred in 57.3 per cent. of the cases, and extension in the remaining 42.7 per cent. When the stimulus was repeated extension was quickly followed by flexion in 75.6 per cent. of the forty-one infants who showed extension in response to a single stimulus. The final response, therefore, was flexion in 89.6 per cent. of the cases. When continued stimulation was applied in cases showing a flexion on the first stimulus, or flexion following extension, no case was observed in which extension followed flexion.

3. *Sticking with a Needle.*—In studying the responses to pain stimuli, the points of stimulation were the cheeks, anterior surface of the thigh, and lateral surface of the calves. The stimulus used was sticking with a needle, the examiner being careful to stimulate with equal intensity in all regions and in all infants. Pulling away of the legs or face associated with movements of the arms and crying were considered a response to the stimulus. The infants were allowed to rest for about fifteen minutes between separate examinations; and when tested were kept awake. When no response was elicited on one stimulus, summation of stimuli was attempted by rapidly repeating the stimulus. The application of more than ten stimuli was not considered feasible.

To stimulation of the legs, three of the four subjects between the ages of 0.5 and 5.5 hours did not react to the greatest number of stimuli. All infants above 76 hours reacted to one stimulus. The number of stimuli necessary to produce a reaction decreased sharply and regularly from 1 to 21 hours, then more slowly to 76 hours. To stimulation of the face, all the subjects tested reacted. The age at which one stimulus was sufficient was much higher for the legs, all subjects more than 41 hours reacting to one stimulus of the face, whereas no subject less than 76 hours of age reacted to one stimulus of the legs.

The difference in threshold between the face and legs may perhaps be accounted for by the fact that the spinal pain fibers have many synapses before reaching the brain, a situation different from that of the fifth cranial nerve. From the functional standpoint, the anterior end of the organism is necessarily much more highly developed, and it is of greater importance for the organism to react to a stimulus of the head end than to one of the caudal end.

4. *Coordination of the Eyes.*—This was tested by moving a flashlight to and fro about 15 inches in front of the infant's face within a range of 1 foot. The number of eye movements observed in each test was limited to fifteen, and the number of coordinated eye movements in the process of fixation was counted.

The ratio of the number of uncoordinated movements to the total number was obtained and the percentage taken as the percentage of error. The results showed that infants more than 34 hours of age showed no error in coordination in the process of fixation on the light. In the youngest subject, aged 2½ hours, the error was 63 per cent. The amount of error decreased fairly regularly with advance in age up to 34 hours. Some of the infants at the earliest ages showed good coordination in one direction, but a large amount of error when changing the direction of fixation on the light. In several cases, it was observed that both eyeballs moved externally. The results indicate that the new-born infant is able to fixate a faint white light, but that the coordination of the eye muscles is poor. With increasing age, the coordination is gradually perfected until the age of 34 hours, when perfect coordination is possible.

5. *Coordination of the Arms.*—This was studied by eliciting defense reactions by pressure on the chin with the examiner's finger, the hand of the examiner being held parallel with the infant's chest so as not to interfere with the movements of the arms. In order to have comparable data, more than thirty movements of the arms were not counted. No infant below 21 hours of age made a perfectly coordinated movement. Between the ages of 21 and 41 hours, all the infants made coordinated movements, the average number of trials necessary being twelve. Between the ages of 41 and 61 hours the average number of arm movements necessary was nine and three-tenths. Above this age group, the decrease was fairly regular. No infant made a coordinated defense movement in less than four trials. (A coordinated movement was considered as one in which both arms touched the distal part of the examiner's finger with a pushing motion.)

Book Review

INTRODUCTION TO THE HISTORY AND HISTOPATHOLOGY OF THE NERVOUS SYSTEM. By Dr. Paul Schröder, Professor of Psychiatry and Neurology in Greifswald. Authorized translation from the second revised German edition by Baldwin Lucke, M.D., Assistant Professor of Pathology, School of Medicine, University of Pennsylvania, and Morton McCutcheon, M.D., Instructor in Pathology, School of Medicine, University of Pennsylvania. Cloth. Price, $3.50. Pp. 161, with 53 illustrations. Philadelphia: J. B. Lippincott Company, 1923.

Dr. Paul Schröder's little book is now available in English, in a faithful translation by Lucke and McCutcheon. Schröder needs but little introduction to neurologists in this country. His book is for the elementary student of neurology, and gives in general terms and in a readable form a bird's-eye view of the changes that may occur in so complex a structure as the brain. It takes up in a routine manner a study of the ganglion cells, neurofibrils, nerve fibers, mesodermal tissue, and the pathologic changes that may occur in these structures. This little book will be of value to teachers and students of neurology.